SPECTRUM OF ORBITAL TUMORS IN A TERTIARY EYE CARE CENTRE

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CERTIFICATE

*Certified that this dissertation entitled "**SPECTRUM OF ORBITAL TUMORS IN A TERTIARY EYE CARE CENTRE** "submitted to the Tamilnadu Dr M.G.R Medical university, Chennai February 2006 is the bonafide work done by **DR.PRASHANT RAPHAEL** under our supervision and guidance in the orbit and occuloplasty department of Aravind eye hospital and post graduate institute of ophthalmology, Madurai during his residency programme from April 2003 to March 2006.*

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INTRODUCTION

The orbit contains variety of tissues including bone, extra ocular muscles, peripheral nerves, cranial nerves, fibro vascular adipose tissue and cartilage and a variety of systemic disorders may manifest in the orbit of which orbital tumors constitute the major category. Diseases of the orbit create some of the most complex and perplexing problems in ophthalmology The initial clinical evaluation of the patient with an orbital lesion is frequently inconclusive .After taking the patient's history and then examining the patient there invariably remains enough uncertainties to require consultation with other specialists and referral for radiological and ultrasonographic studies . Very often, a desirable end result of the workup is the localization of the lesion to a particular area of the orbit, thereby facilitating the surgical exposure of the lesion and permitting its excision. Such a complex subject deserves a precise table of organization; unfortunately none exists. The small confines of the orbit and the anterior location of the eye ball limit direct observation or palpation of orbital tumors. Even knowledge of the approximate size, location and consistency of the lesion is not effective means of sorting out the various lesions of the orbit. Additional information regarding the direction degree and rate of progression of proptosis and other information

does not facilitate the formulation of a workable classification. Classifications of the orbital disease derived from the use of ultrasonography and computerized tomography have become very helpful; however such data are usually not available at the initial evaluation. Perhaps the most commonly used classification is one based on the review of histopathologic material [1]. The reported incidence of orbital tumors varies widely from series to series depending on the material studied .One finds a considerable difference in the incidence of various lesions in series derived from biopsy material as compared with those that include clinical diagnosis without histopathological verification .Series from large ophthalmic departments have inherent bias depending on the referral patterns special interests of the ophthalmologist and other factors [2]. This study intends to study the spectrum of orbital tumors in a tertiary eye care centre.

REVIEW OF LITERATURE

The reported incidence of orbital tumors varies from series to series depending primarily on the source of the material reviewed .Variation in frequency of different tumors in different series can have relationship to national ,regional and other socioeconomic factors. Biopsy proven series is recognized for the certainty and prognostic aspect. A clinical series represents a wider practical correlation and may include cases not likely to be biopsied.

Daniel Silva M.D published a study in 1968 on orbital tumors which included 300 patients, all of whom underwent surgery at least for biopsy .Pseudo tumors were the most common group followed by mucocoeles and orbital expansion of retinoblastoma. The unusually high incidence of retinoblastomas with orbital involvement was explained by the fact that the hospital was an oncology hospital where only children referred for radiotherapy or chemotherapy were seen.[3]

O.P.Kureshtra et al presented a study consisting of 104 cases seen over the last 20 years .Benign primary tumors consisted 48 ,malignant (primary)22, malignant (metastatic)3,extensions from paranasal sinuses malignant 25,benign 1,extension from the cranial fossa .[4]

Robert Kennedy reviewed 820 clinical orbital cases over a 34 year period, which include biopsy proven (450) and other clinical cases .These were patients seen by ophthalmologists practicing in an average community area rather than a large referral center and thus may be truly representative of what might be encountered in practices. This study included cases which were biopsy proven and also cases which were diagnosed without biopsy when there was strong medical, xray, or computerized tomography evidence Trauma, lymphoma, dermoid and idiopathic orbital inflammation were the most common diagnoses. [5]

Jerry Shields et al reviewed 645 consecutive biopsies of orbital lesions performed at a major ophthalmic hospital during a 20 year period .Their series of 645 specimens included dermoid, dacryoadenitis, reactive lymphoid hyperplasia and lymphoma as the most common lesions. This study included a group of primary orbital melanocytic tumors which were absent in the other studies.[6]

Between 1977 and 1984, 84 cases of orbital tumors with histological basis were analyzed at Chang Gung memorial hospital, Taiwan. There were cases with primary orbital tumors (70.2%), secondary orbital tumors (27.4%), and metastatic orbital tumors (2.4%).The most common primary orbital tumor was the dermoid cyst and hemangioma .Adenexal squamous cell carcinoma, retinoblastoma, and mucocoele were the secondary tumors .Nasopharyngeal cancer in secondary orbital tumor was also common.[7]

Vijayalakshmi Ammal reviewed 212 cases of orbital tumors of which 84 cases were neurofibromas and 64 were lacrimal gland tumors in 1990.[8]

Sylvia et al reviewed cases of histopathologically verified orbital tumors in children to

determine the distribution of various pathological processes and trends over time .The medical records and pathology specimens from 340 patients aged 18 years or younger who under went biopsy from orbital mass in 1932 to 1991 were reviewed. The most common tumors were cysts, vascular lesions optic nerve and meningeal neoplasms. The overall frequency of malignancy was 18.2%of which 11.5%were primary tumors and 6.8 % were secondary. [9]

Henderson summarized his clinical experience at the Mayo clinic .He had a total of 1376 cases over a time period of 18 years ,and his cases were from surgically excised specimens and angiographic findings in case of vascular malformations. Mucocoele, lymphoma, squamous cell carcinoma, and meningioma were the most frequent lesions in his review.[10]

Wilson et al reviewed 312 orbital lesions accessioned in their laboratory between 1942 and 1993 .Orbital invasion of uveal melanoma, idiopathic orbital inflammation and dermoid were the most common lesions in their series.[11]

Demirci, Hakan et al in 2002 reviewed approximately 950 cases of orbital space occupying lesions in the older adult population. The orbital tumor was unilateral in 183 patients and bilateral in 17 patient's .The most common clinical features included mass proptosis and pain .The most common diagnosis were malignant lymphoma, idiopathic orbital inflammation and cavernous hemangioma.[12]

Jerry shields et al reviewed 1264 consecutive patients with orbital tumors and stimulating lesions referred to an ocular oncology center .In this series 64% were benign and 36% were malignant. The most common diagnosis was lymphoid tumor, idiopathic orbital inflammation and cavernous hemangioma.[13]

PERSPECTIVE OF ORBITAL TUMORS

CLASSIFICATION OF ORBITAL TUMORS

CLASSIFICATION OF ORBITAL TUMORS

According to Grossniklaus, the orbital disorders can be classified into (I) Systemic (II) Inflammatory (III) Trauma (IV) Congenital (V) Primary neoplasms (VI) Secondary neoplasms (VII) Metastasis (VIII) Vascular (IX) Others. Tumors were classified into primary, secondary and metastatic. Primary tumors were further classified into [2]

- *EPITHELIAL*
  - BENIGN MIXED TUMOR
  - MALIGNANT MIXED
  - ADENOID CYSTIC CARCINOMA
  - SPINDLE CELL CARCINOMA
  - ADENO CARCINOMA
  - MUCOEPIDERMOID CARCINOMA
  - LYMPHOEPITHELIOMA
  - CONJUNCTIVAL CYST
  - SUDORIFEROUS CYST
- *FIBROUS CONNECTIVE TISSUE*
  - FIBROMA
  - FIBROSARCOMA
  - FIBROUS HISTIOCYTOMA
  - MYXOMA
  - FIBROMATOSIS

- ***FIBROOSSEOUS***
  - OSTEOMA
  - OSTEOSARCOMA
  - FIBROUS DYSPLASIA
  - OSSIFYING FIBROMA
  - PAGET'S DISEASE
  - ANEURYSMAL BONE CYST
  - FIBROOSTEOMA
  - EWING'S SARCOMA
- ***CARTILAGENOUS***
  - CHONDROMA
  - CHONDROSARCOMA
- ***ADIPOSE***
  - LIPOMA
  - LIPOSARCOMA
- ***VASCULAR***
  - CAPILLARY HEMANGIOMA
  - CAVERNOUS HEMANGIOMA
  - LYMPHANGIOMA
  - HEMANGIOPERICYTOMA
  - HEMANGIOENDOTHELIOMA
  - ANGIOLEIOMYOMA
  - FIBROANGIOMA

- GLOMANGIOMA

❖ *MUSCLE*

- RHABDOMYOMA
- RHABDOMYOSARCOMA

❖ *NEURAL*

- MENINGIOMA
- NEUROFIBROMA
- GLIOMA
- NEURILEMMOMA
- MALIGNANT PERIPHERAL NERVE SHEATH TUMOR
- ASTROCYTOMA
- EPENDYMOMA
- GRANULAR CELL MYOBLASTOMA

❖ *MALIGNANT LYMPHOMAS*

- LYMPHOID HYPERPLASIA
- WELL DIFFRENTIATED LYMPHOCYTIC TYPE
- POORLY DIFFRETIATED LYMPHOCYTIC TYPE
- HODGKINS DISEASE
- RETICULUM CELL TYPE

❖ *OTHER TUMORS OF RETICULUM CELL PROGENY*

- HISTIOCYTOSIS
- INFANTILE XANTHOGRANULOMA

- PLASMA CELL DYSCRASIAS
- LEUKEMIA

SECONDARY NEOPLASMS

- INVADE THE ORBIT FROM ADJACENT STRUCTRES INCLUDING THE EYE AND OCULAR ADENEXA, PARANASAL SINUSES, ORAL CAVITY AND CRANIUM.

METASTATIC TUMORS

- TUMORS METASTAZISING TO THE ORBIT

ORBITAL TUMORS

VASCULAR

CAPILLARY HEMANGIOMA[23]

DEFINITION: Common benign hamartoma (mass of disorganized but mature specialized cells or tissue indigenous to the particular site) of orbital & periorbital areas in childhood. **INCIDENCE:** 1-2% of orbital tumor. **AGE/SEX:** Usually presents in first few months of life (95% by 6 months of age), but never at birth with female preponderance. **CLINICAL FEATURES**: Mainly of 3 types being Superficial type presenting as "strawberry nevus", deep variety in orbit giving rise to proptosis and combined variety. **HISTOLOGY:** It is composed of endothelial & capillary vessel proliferations with benign endothelial cells surrounding the small capillarised vascular spaces. **RADIOLOGY**: On CT-Scan, appears fairly well marginated or poorly marginated, irregular enhancing lesions. On MRI, appears hypo or slightly hyper intense to brain in T1 weighted images and hyper intense in T2.**MANAGEMENT**: Indicated in cases of amblyopia, optic nerve compression, exposure keratitis or cosmetic blemish with necrosis. It includes intralesional steroids, systemic steroids, alpha-interferon, local resection & low dose radiotherapy.

CAVERNOUS HEMANGIOMA[24]

DEFINITION: Type of developmental polymorphic vascular hamartoma commonly seen in adults. . Most are located within the muscle cone.

INCIDENCE: 1-2% of orbital tumour.**AGE GROUP**: 1st to 8th decade. Mainly in middle aged women (M: F=1:3) .**CLINICAL PRESENTATION:** Unilateral, painless, gradually progressive axial proptosis usually asymptomatic. Can cause decreased vision due to optic nerve compression or induced hyperopia, amaurosis due to intermittent optic nerve compression. **HISTOLOGY:** Well-circumscribed, well encapsulated, mulberry like, spongy , reddish brown colored mass On cut-section —large blood-filled spaces. **RADIOLOGY:** CT-scan: Well-defined, rounded, intraconal mass with smooth margin, with uniform homogenous enlargement on contrast. They do not deform the globe. Serial dynamic MRI shows connecting point of tumor to the feeder vessel in early stage (50 sec). In T-1image: iso intense with muscles. In T-2 image, hyper intense to fat and muscles.
MANAGEMENT: Surgical excision in symptomatic patients and the common site being intraconal, lateral orbitotomy is the surgery of choice.

LYMPHANGIOMA[25]

DEFINITION: Benign vascular malforma tions, which are abortive and nonfunctional. . This lesion is seen most frequently in young children.This lesion may wax and wane in size, especially when the child has an upper respiratory infection(reflecting the proliferating lymphoid elements in the connective tissue trabeculae of the tumor). **INCIDENCE:** Less than 7% of patients with childhood orbital tumors **AGE/SEX**: Mainly in 1st two decades of life, M: F=1:3.**CLINICAL FEATURES**: If anterior in orbit, multiple small bluish masses in superonasal quadrant of orbit. If posterior, it causes slowly progressive proptosis ,which spontaneously become painful, due to hemorrhage in tumor it will form "chocolate cysts" which will resolve with time. **MICROSCOPY:** Non encapsulated networks of endothelially lined channels, separated by thin septae and filled with pale-staining lymph. **RADIOLOGY**: Poorly circumscribed heterogeneous masses of increased density in extra & intraconal space. Fluid-fluid levels related to hemorrhages of various stages are best seen in MRI **MANAGEMENT:** Persistent sight-threatening chocolate cysts should be drained or removed sub totally by controlled vaporization of CO2

laser, otherwise conservative treatment is gold standard.

HEMANGIOPERICYTOMA/HEMAGIOENDOTHELIOMA[26]

DEFINITION: Group of tumors with rich vasculature. In Haemangiopericytomas, where capillaries consisting of endothelial channels surrounded by proliferation of contractile pericytes of Zimmerman. In Haemangioendothelioma there are both locally aggressive and capable of metastasis where it is called malignant haemangioendothelioma.**INCIDENCE:** About 15-30% of hemangiopericytomas occur in the head and neck region. haemangioendothelioma compose less than 1% of all soft tissue sarcoma .**AGE/SEX**: The tumor can appear in persons of any age, with 80-95% of patients older than 20 years.). Approximally 10% of hemangiopericytomas are found in children .Males=Females.**CLINICAL PRESENTATION**: About 50% cases are malignant having potential for distant spread. In orbit, generally presents as non tender, irreducible and non-pulsatile proptosis, superior orbit and intraconal location being common. Capable of metastasizing to distant organs .In striking contrast to cavernous hemangioma, these cause puffiness of the eye lids with bluish discoloration of the ocular adenexa.**PATHOLOGY:** Histopathologically it may assume one of 3 configurations:1)A prominent vascular pattern of sinusoidal spaces,2)An ostensibly solid pattern in which the vasculature may not be seen.3)A combination of the two. Vascular pericytes are of mesenchymal origin that spiral around capillaries and postcapillary venules. The tightly packed cells around the thin-walled blood vessels are lined by flattened endothelial cells. Hemangiopericytomas contain periodic acid schift and reticulin-staining basement membrane material.**RADIOLOGY**: Orbital CT Scan

documents a round to oval mass usually located in the superior orbit.The well-encapsulated lesion,with its large arterial feeding vessels,exhibits dramatic enhancement on injection of contrast material. Slightly less distinct margins is due to invasion of adjacent tissues.Erosion of underlying bone and marked contrast enhancement are key to diagnosis.On MRI ,these well-delineated tumors are hypointense to fat on T1-weighted images and hyperintense to fat and isointense to vitreous on T2-weighted images. **MANAGEMENT**: Optimal treatment is wide excision with negative margins. Tumor cells may be left behind with enucleation. For unresectable or incompletely excised tumors, radiotherapy has an important role.

FIBROUS CONNECTIVE TISSUE

FIBROMA[28]

DEFINITION: It is a true benign neoplasm of bone originating in the mesenchymal tissue of periosteum. **INCIDENCE:** 4%-6% **AGE/SEX:** Younger patient,7-18 years (average 13) **CLINICALPRESENTATION:** Slowly progressive painless proptosis. Orbital plate of frontal and spheno ethmoidal bone commoly affected. It consists of units of lamellar bone which show clear-cut lines or lamellae on polarization. Osteoblastic activity is present.**HISTOPATHOLOGY:** Usually tan-gray colored whitish red tissues with soft fibrous texture having grittiness depending on osteoid formation. Highly cellular fibroblastic stroma interspersed with bony spicules forming osseous trabeculae rimmed by osteoid which is again rimmed by osteoblasts.Typically shows ”zonation phenomenon” showing more mature osteoids towards periphery.**RADIOLOGY:** CT shows a mass with less radiodense centre with scattered calcifications with sclerotic margin, more delimited than fibrous dysplasia. **MANAGEMENT:** Wide local excision.

FIBROUS HISTIOCYTOMA[29]

DEFINITION- Malignant fibrous histiocytoma (MFH), described by O'Brien and Stout in 1964, is the most common soft tissue sarcoma of late adult life due to fibroblastic proliferation involving fascia, muscles and soft tissues .**INCIDENCE**- MFH accounts for 20-24% of soft tissue sarcomas, making it the most common soft tissue sarcoma occurring in late adult life.**AGE/SEX**- The tumor occurs with a peak incidence in the fifth and sixth decades but an age range of 10-90 years is reported. Although the

tumor is rare in children, the angiomatoid subtype is the most frequently occurring variety in patients younger than 20 years. Male-to-female ratio is approximately 2:1. **CLINICAL PRESENTATION**-Slowly growing proptosis with growth in superonasal quadrant of orbit, with congestion, chemosis, lid retraction, motility disturbances and occasionally compressive neuropathy. **PATHOLOGY:** The tumor contains both fibroblast like and histiocyte like cells in varying proportions, with spindled and rounded cells exhibiting a storiform arrangement. Five histologic subtypes have been described including (1) storiform/pleomorphic (most common), (2) myxoid, (3) giant cell, (4) inflammatory (usually retroperitoneal), and (5) angiomatoid (often located more superficially than other varieties).**RADIOLOGY**-CT shows well-circumscribed mixed density tumor with heterogeneous enhancement and intact bones. USG reveals a high tissue reflectivity and heterogeneity. Benign lesions will show compressive bone remodeling. **MANAGEMENT**-Benign-surgical excision. Malignant-Exentration and radiotherapy /chemotherapy.

OSSEOUS

OSTEOMA[30]

DEFINITION: Osteoid osteoma is a benign skeletal neoplasm of unknown etiology that is composed of osteoid and woven bone. **INCIDENCE:** Accounted for 12.1% of benign tumors and 2.9% of all tumors 10%-15%**AGE/SEX:** Three quarters of patients are in age 10-30 years, and more than 90% of patients are aged 5-25 years. The age range in patients is 5-56 years. The male-to-female ratio is 2:1. **CLINICAL PRESENTATION:** Focal skeletal bone pain, which worsens at night and is frequently relieved with a small dose of aspirin. Nonaxial Proptosis. Frontal and ethmoidal sinuses involvements are the most common presentation in and around orbit. **PATHOLOGY**: The tumor consists of an ovoid or spherical nidus of osteoid-rich tissue and interconnected bone trabeculae superimposed on a background of highly vascularized connective tissue containing large dilated vascular channels. **RADIOLOGY:** CT shows hyperdense, rounded or multilobular lesion projecting into the orbit. **MANAGEMENT:** Surgical excision. **ASSOCIATIONS:** Turcot's syndrome , Gardner's syndrome.

CHONDROMA[31]

DEFINITION: Enchondromas are benign cartilaginous neoplasms in bone. . In orbit it envolves trochlear cartilage. **INCIDENCE:** Enchondromas account for 12-14% of benign bone neoplasms and 3-10% of osseous neoplasms in general. **AGE/SEX**: Solitary enchondromas most often are discovered in those aged 20-40 years. Enchondromas occur equally in males and females.**CLINICAL PRESENTATION:** Slowly progressive painless proptosis. Typically located in superonasal quadrant and attached to medial orbital wall. **HISTOLOGY:** Enchondromas are ectopic hyaline cartilage rests in intramedullary bone hypocellular population of cells of hyaline cartilage that are widely dispersed singly in lacunae. **RADIOLOGY:** The lesions replace normal bone with mineralized or unmineralized hyaline cartilage, thereby generating a lytic pattern on radiographs or a lytic area containing rings and arcs of chondroid calcifications. **MANAGEMENT**: Wide local excision

OSSIFYING FIBROMA[28]

DEFINITION: Osteofibrous dysplasia which occurs exclusively in adults commonly is referred to as ossifying fibroma. **INCIDENCE**: Osteofibrous dysplasia usually is first diagnosed in children with a peak incidence occurring in children aged 1-5 years. **AGE/SEX:** The age range at the time of

diagnosis has been variable in the literature.No significant sex preponderance consistently has been reported. **CLINICAL PRESENTATION**: Slowly progressive painless proptosis. Orbital plate of frontal and sphenoethmoidal bone most commonly affected.

PATHOLOGY: Highly cellular fibroblastic stroma containing interspersed with bony spicules forming osseous trabeculae rimmed by osteoid which is again rimmed by osteoblasts. Typically shows,"Zonation phenomenon" showing more mature osteoids towards periphery.**RADIOLOGY**: Osteofibrous dysplasia is an eccentric, intracortical, osteolytic lesion. CT shows a mass with less radiodense centre with scattered calcifications with sclerotic margin..**MANAGEMENT**: Due to the high recurrence rate, nonoperative treatment of the lesion until after skeletal maturity is reached, at which time marginal resection and bone grafting may be performed without increased risk of recurrence.

MUSCLE

RHABDOMYOSARCOMA[32]

DEFINITION: Rhabdomyosarcoma (RMS) (from Greek, *rhabdo,* meaning rod shape, and *myo,* meaning muscle) is the most common soft tissue sarcoma in children which arises from undifferentiated mesenchyme rather than differentiated striated muscle.Pre-existent radiotherapy, germ line mutation of p53 suppressor gene(Li-Fraumeni Syndrome), translocation (2;13), (q35;q14).**INCIDENCE:** 6:1,00,000 per year. 5% of childhood malignancies, 50% of all paediatric sarcomas and 15% of all paediatric solid tumors. 44% in head and neck- 25%-60% of these originate in orbit. **AGE/SEX:** Approximately 87% of patients are younger than 15 years, and 13% of patients are aged 15-21 years. RMS rarely affects adults. In patients with orbit RMS, 42% are aged 5-9 years Bimodal age distribution, with higher .prevalence in children, average age being 7 years (Embryonal & Alveolar) whereas Pleomorphic type is common in adults. Overall, the male-to-female ratio is 1.2-1.4:1. Differences exist according to the site of primary disease.In orbit: The male-to-female ratio is 0.88:1. **CLINICAL PRESENTATION:** Acute onset of painless, rapidly progressive proptosis. Swelling and ptosis of upper lid caused by a palpable mass usually in upper quadrant. **PATHOLOGY**: RMS is one of the small, round, blue cell tumors of childhood. RMS cells may demonstrate positive immunohistochemistry results for muscle-specific markers, such as myoglobin, actin, and desmin. Cells from the RMS subtypes have the following distinctive features: (1) Botryoid: The cambium layer is characteristic, containing a condensation of loose tumor cells below an epithelial surface. (2) Alveolar: Cells line up

along membranes that may be imperceptibly thin or may be obvious collagen bands, resembling the lung alveoli. Alveolar has worst prognosis (3) Undifferentiated: Usually, no evidence of myogenesis differentiation is present. **RADIOLOGY**: CT shows enhancing mass within the orbit with bone destruction in larger tumors. Its usually isointense in relation to normal muscles, appears homogenous, well-defined soft tissue masses without bone damage. MRI: T1 weighted appears isointense or slightly hypointense compared to brain and appears hyperintense on fat suppression. T2 images shows increased signal intensity.**MANAGEMENT:** Incision biopsy followed by chemotherapy and /or radiotherapy.

NEURAL

MENINGIOMA[33]

DEFINITION: Tumor arising from meningothelial cap cells of arachanoid matter of optic nerve. **INCIDENCE:** It is approximately 20% of all intracranial tumors; population-based studies indicate an overall incidence of 2.3 cases per 100,000.**AGE/SEX:** In children, the male-to-female ratio is 2:1, with an average age at presentation of 10.1 years (range 1-16 y). The incidence of meningiomas increases with age, 2-7 cases per 100,000 in women and 1-5 cases per 100,000 in men. Peak incidence is in the seventh decade in women and in the eighth decade in men. Meningiomas rarely occur in infants.**CLINICAL PRESENTION**: Profound visual loss evolves over decades & early loss apparent only with threshold perimetry. Occasionally patient may present with proptosis. **MICROSCOPY**: The macroscopic appearance of meningiomas may be hemispheric, bun-shaped, or globular. They usually are attached to the dura and invaginate into adjacent neural structures. Enveloped in a thin capsule derived from the adjacent meninges, they remain extraaxial and are separated easily from the brain or the spinal cord. **RADIOLOGY:** CT shows two morphologic types: En plaque- a focal exophytic tumor along the course of intraorbital segment with enhancement. Fusiform- tumor is denser than normal nerve & shows “rail-road track” sign produced by calcification. ONSM is confined to the dura mater; therefore, it often appears as a distinct, fusiform thickening of the optic nerve. This enlargement may appear as localized or as an eccentric expansion of the optic nerve, and it occurs most commonly at the orbital apex.On MRI, tumor is hyperintense on T1 & T2 weighted images. Although an MRI most readily shows the soft tissue tumor characteristics of ONSM, a CT scan better displays any associated bony hyperostosis.**MANAGEMENT:** Patients with ONSM can be observed if there is no evidence of intracranial extension, vision loss, or visual field loss. Treatment with primary radiation or radiation following surgical removal has been associated with a better chance of visual improvement. Chemotherapy is reserved for patients with unresectable, recurrent, or previously irradiated meningiomas. Combination treatment with 5-fluroouracil, folate, and levamisole, or a combination of intraarterial cisplatin with intravenous doxorubicin, may be beneficial.

OPTIC NERVE GLIOMA[34]

DEFINITION: They are juvenile pilocytic astrocytoma, fusiform, ordinarily benign enlargement of optic nerve contained within an intact dural sheath**. INCIDENCE :** Optic nerve gliomas represent 4% of orbital tumors, 4% of intracranial gliomas, 2% of intracranial tumors and two thirds of all primary optic nerve tumors.**AGE/SEX**: In the pediatric population, the median age of patients is 5 years, and 80% of patients present prior to age 15 years . In adult patients, age ranges from 22-79 years, with a mean age of 52 years . In pediatric patients, a slight female predominance is found, while in adult patients, a slight male predominance is found.**CLINICAL FEATURES**: The presenting symptom is painless proptosis. Optic atrophy is common When the lesions are large, local compressive effects may

result in optic chiasm symptoms such as nystagmus. Hypothalamic symptoms, such as changes in appetite or sleep, also may occur. When lesions are massive, compression of the third ventricle may occur,resulting in obstructive hydrocephalus with headache,nausea, and vomiting.**HISTOPATHOLOGY:** consists of spindle cells with frequent mucinous changes.rosenthal fibres are pathognomic with fibrillary astrocytes positive for phosphotungstic acid hematoxylin and glial fibrillary acidic protein. **RADIOLOGY:** In children, unenhanced CT typically reveals a marked diffuse enlargement of the optic nerve, with characteristic kinking or bending. The enlargement may be tubular, fusiform, or excrescent. Areas of lucency may be observed as a result of mucinous or cystic changes. Approximately 50% of the lesions demonstrate enhancement, and enhancement is more common with intracranial (especially retrochiasmatic) extension. Calcifications are rare. MRI shows hypotense or isotense lesion on T1 & hyperintense on T2 weighted images. Enlargement of optic canal. MRI shows hypotense or isotense lesion on T1 & hyperintense on T2 weighted images. Intracranial extension better appreciated on MRI. **MANAGEMENT:** Guided by location of the tumor & visual potential. Usually observation with monitoring of optic nerve function & disc edema 3 monthly .Surgery is the main stay of treatment.

NEUROFIBROMA[35]

DEFINITION: Orbital neurofibroma is usually an isolated tumor of nerve cells, and is not associated with neurofibromatosis. Orbital neurofibroma that is associated with neurofibromatosis also occurs, and this type is typically the plexiform neurofibroma, which begins in the eyelid. Consits of a mixture of Schwann's cells, peripheral nerve axons, endoneural fibroblasts & perineural cells. ssociated with neurofibriomatosis-1 which is an autosomal dominant inherited trait. **INCIDENCE**: 0.5%-2.4% of orbital tumors. **AGE/SEX:** Localised neurofibroma in young to middle-aged patient and diffuse and plexiform neurofibroma make their appearance within first decade of life.**CLINICAL PRESENTATION:** Three types**:** Isolated neurofibromas, plexiform, diffuse. Isolated NF presents a painless of mildly painful proptosis caused by an orbital soft tissue mass in superior quadrant commonly. It's a non-encapsulated-has compression pseudocapsule-white to tan colored tumor. In Plexiform and Diffuse NF patient presents with massive overgrowth of lids along with similar infiltration of orbit and facial tissues, In the first there is vermiform collection of molluscusm,Lisch's nodule,axillary freckle etc. In Plexiform, tissues are expanded by recognizable neural units whereas Diffuse form observes a monotonous spindle cells proliferation. **MICROSCOPY**: Consists of wavy cells typically dispersed in small bundles. In Plexiform and Diffuse neurofibromas, patient presents with massive overgrowth of lids along with similar infiltration of orbit and facial tissues, in the first there is vermiform collection of individual units on palpation not in the later. Other features are café-au-lait spots, fibroma molluscusm, Lisch nodule, axillary freckling. In Plexiform, tissues are expanded by recognizable neural units whereas diffuse form observes a monotonous spindle cell proliferation. **RADIOLOGY:** In isolated form CT reveals a homogenous well-circumscribed soft tissue density that has uniform contrast medium enhancement. MRI shows lesions hypointense to isointense on T1 and hyperintense on T2 weighted images. In plexiform neurofibromas, CT shows moderately enhancing soft tissue density without clearly defined borders. May show absence of sphenoid wing..**MANAGEMENT**: Surgical excision in localized form. In plexiform and diffuse , cosmetic debulking using CO_2 laser.

NEURILEMOMA[36]

DEFINITION: A pure proliferation of Schwann's cells. Orbital schwannomas are usually isolated lesions, except when they are associated with neurofibromatosis type 2 (NF2), a rare autosomal dominant disorder occurring in approximately 1 live birth in 50,000. NF2 is also called the multiple inherited schwannomas, meningiomas, and ependymomas (MISME) syndrome. It arise from the nerve

sheath ,consist of Schwann cells in a collagenous matrix.**INCIDENCE:** Schwannomas account for 6-8% of intracranial neoplasmsAccounts for 1% of all orbital tumors. Are found in 1.5% of patients with NF1. **AGE/SEX:** Young to middle aged adults. No sex predilection has been described.**CLINICAL PRESENTATION:** Produce slowly progressive painless proptosis. **HISTOPATHOLOGY:** They are extremely encapsulated, have smooth surface & friable. Usually appears yellowish-tan colored, solid, encapsulated with varicose, violaceous tumor vessels on surface. They have both solid and myxoid areas alternate with each other in varying proportion. Spindle-cells oriented in fascicles, with long axis parallel to each other called Antoni Type A., in type B it is haphazardly arranged. **RADIOLOGY:** CT shows a homogenous well-circumscribed soft tissue mass. On nonenhanced CT scans, most schwannomas are isoattenuating relative to brain parenchyma. Calcification or areas of hemorrhage are rare. Long standing lesions may produce remodeling or frank erosion of bone. MRI shows a discrete,homogenous soft tissue density that is hypointense on T1 and hyperintense on T2 weighted images. The cystic spaces can result in high signal intensity on T2-weighted MRIs. Gadolinium enhancement is typically homogeneous, although larger schwannomas can show areas of cystic degeneration and heterogeneous signal intensity; these findings are based on increased numbers of areas with Antoni type B histologic features. **MANAGEMENT:** Stereotactic radiosurgery (ie, gamma knife radiosurgery) largely has replaced surgical resection for the treatment of vestibular schwannomas, particularly when the lesions do not compress the brainstem. Lesions should be smaller than 3 cm. Studies have demonstrated rates of tumor control (ie, lesion stabilizes or shrinks) of greater than 95% and a rate of hearing preservation of approximately 70%. Although less well studied, other CN schwannomas also can be treated with radiosurgery

MALIGNANT LYMPHOMAS[37]

DEFINITION : These are solid tumors of immune system composed of monoclonal B-cells. It mimics Non-Hodgkin's lymphoma. The lacrimal gland is the most frequent site of involvement in the orbit. It may be either due to systemic disease(75%) or may indicate primary site. Bilateral involvement can also be seen. 2 parts in orbit having lymphoid tissue, i.e. substantia propria of conjunctiva & lacrimal glands,account for lymphoreticuloses of orbit .Of all the patients of orbital lymphoma,75% will have systemic disease. **INCIDENCE**; Almost up to 10-15% of orbital masses are extranodal NHL.**AGE/SEX**: Rarely in children.Mainly in 6-7 decade of life with slight female preponderance. **CLINICAL PRESENTATION:** They usually occur in anterior orbit with pink, fleshy, sub conjunctival tumefaction (salmon patch), tends to mould to the shape of globe. On palpation they are nodular with well-defined margins and slightly

friable texture with rich vascularity intra operatively. Roughly 75% lesions are unilateral. It produces painless progressive non axial proptosis, visual disturbances & lacrimal gland enlargement Any pt of orbital lymphoma should be subjected to systemic evaluation to rule out systemic lympho proliferative disorder. **PATHOLOGY:**The cells are monotonous with a high nuclear-to-cytoplasmic ratio.Nuclei often are convoluted with finely stippled chromatin; nucleoli usually are visible but not prominent. Immunohistochemical analysis reveals T-cell markers,including CD5 and CD7.**RADIOLOGY:** They are homogenous masses with relatively high density & sharp margins, mainly in anterior orbit, superior orbit or retrobulbar area. It shows putty like molding of tumor to preexisting structures without eroding the bone or expanding the orbit. Lacrimal gland lymphoma displaces the globe medially & forward. The optic nerve lymphomatous infiltration shows tram-like (axial) and ring (coronal) enhancement after IV contrast which is also noticed in meningiomas and sarcoidosis. **MANAGEMENT:** Low dose radiotherapy and chemotherapy for systemic cases.

TUMORS OF RETICULUM CELL ORIGIN[37]

Histiocytosis X or Langerhans' cell histiocytosis

DEFINITION + CLASSIFICATION: A group of histiocytic proliferative disorders derived from dendritic types of cells derived from bone marrow, epidermis, brain & oral mucosa.Mainly classified into 3 groups [1].Eosinophilic granuloma (self limited, benign) [2].Hand-Schuller-Christian Disease (Multi focal with systemic involvement) [3].Letterer-Sewe Disease(Acute , disseminated disease).**INCIDENCE:** Comprises 7% of orbital tumors in children and <1% of total orbital tumors. **AGE/SEX** : Mainly in early childhood , 1 to 3 years of age with M:F=3:1.**CLINICAL PRESENTATION:** [1]. Eosinophilic granuloma - comprises 70% of all histiocytosis, mainly solitary, orbital mass with painful ,tender, erythematous swelling at anterior & superotemporal orbit causing proptosis. Skull bones are the commonly involved with frontal & parietal being commonest. [2]. Hand-Schuller-Christian Disease - comprises 20% of all histiocytosis, with equal sex distribution. It presents as multifocal bony lesions with hepatosplenomegaly, lymphadenopathy, malaise, anorexia, fever & classical triad of exophthalmos, diabetes insipidus & bony destructions.[3]. Letterer-Sewe Disease —

comprises 10% of histiocytosis, with acute, disseminated histiocytosis in infants<2yrs.It shows hemorrhagic papules of skin, spleenic & hepatic involvement, pancytopenia with poorest 5 year survival (<50%).Orbital involvement is very rare here. **HISTOPATHOLOGY:**Usually the tumors are soft, friable, hemorrhagic & tan-yellow colored. Microscopy shows large Langerhans histiocytic cells (15-25 microns) with round to indented nuclei and marked nuclear folds resembling coffee beans. Typical rod-shaped, tubular, tennis racquet shaped cytoplasmic inclusions called Birbeck granules are diagnostic on electron microscopy. Immuno histochemical markers are nuclear membrane S-100 protein & cell-surface and cytoplasmic marker called CD1a (OKT-6). **RADIOLOGY**: Bone involvement is characterized by irregular lucent patches with cortical destruction but without any sclerosis. In orbit mainly the superotemporal part shows pathology. **MANAGEMENT:** For localized disease, localized curettage with intralesional steroids followed by low dose radiation and systemic steroids. For disseminated diseases, chemotherapy is a must.

Granulocytic sarcoma (Chloroma)

DEFINITION: Extra medullary form of Acute Myeloblastic Leukemia **INCIDENCE:** It occurs in about 3% of all AML patients.**AGE/SEX:** Mean age of onset is 7 years with Male preponderance (3:2) and non-Caucasian population. **CLINICAL FEATURES:** As orbit being the site of initial presentation showing involvement of soft tissues, lacrimal glands & bones. The lesion typically involves subperiosteal space of lateral orbital wall with extension into temporal fossa. **HISTOPATHOLOGY:** Typical greenish hue of tumor is due to enzyme of tumor cells. Cells of myeloid origin are best seen by Leder stain for chloroacetate esterase. **RADIOLOGY:** Depending on area of infiltration, mainly subperiosteal space of superotemporal quadrant with proptosis. **MANAGEMENT** Radiotherapy and chemotherapy.

INTRINSIC NEOPLASMS OF THE LACRIMAL GLAND[38]

PLEOMORPHIC ADENOMA

DEFINITION: Is a benign mixed-cell tumor and most common epithelial tumor of lacrimal gland. It is derived from ducts, stroma, and myoepithelial elements.**INCIDENCE:** It is the most common epithelial tumor of lacrimal gland. Epithelial neoplasms account for only 4% of all lacrimal gland lesions. **AGE/ SEX:** Second to Fifth decade of life. M=F.**CLINICAL PRESENTATION:** Slowly progressive swelling in upper outer quadrant of more than 1 year duration and may cause non-axial proptosis. **HISTOPATHOLOGY:** firm, grayish-white bosselated and solitary masses. Mixture of epithelium and connective tissue with pleomorphism. Pseudocapsule is present showing microscopic nodular extensions into pseudocapsule in 60% cases. **RADIOLOGY:** CT shows a round or oval smooth outline with excavation of lacrimal gland fossa with destruction with moderate to marked enhancement on CT and MRI.**MANAGEMENT:** Complete surgical excision without violating the capsule. Incisional biopsy of these lesions is contraindicated because, although histologically benign, incomplete excision often leads to repeated recurrences (as high as 30% in some studies) and malignant transformation.

ADENOID CYSTIC CARCINOMA

DEFINITION: Most common malignant epithelial tumor of lacrimal gland. **INCIDENCE:** Adenoid cystic carcinoma is the most common malignant lacrimal gland tumor, comprising 50% of malignant tumors of lacrimal gland and 25% of all lacrimal gland-tumors.**AGE/SEX**: 4^{th} to 6^{th} decade.**CLINICAL**

PRESENTATION: Proptosis of shorter duration (<10 months) and with pain associated with parasthesiae. **HISTOPATHOLOGY:** Derived from duct cells, and they form spaces into which basement membrane like material is deposited. This confers a cribriform or "Swiss cheese" appearance to the tissue. Five histologic patterns have been observed in these lesions, as follows: (1) cribriform (the most common subtype) (2) sclerosing, (3) basaloid, (4) comedo, and (5) ductal. The basaloid type has the worst prognosis.**RADIOLOGY:** CT shows large tumors, calcification with bony erosions having destruction of walls and pressure changes having expansion and lytic lesions of lacrimal fossa with micro serrations. MRI is useful to see perineural infiltration, cavernous sinus involvement.**MANAGEMENT:** Wide local excision followed by radiotherapy or brachytherapy

SECONDARY NEOPLASMS[39]

DEFINITION: Involves the orbit by direct extension and can arise in any of adjacent structures. Para nasal sinus, Nasopharyngeal, Lacrimal, Conjunctival, Eyelid, Intraocular, Intracranial tumors. **INCIDENCE:** 48%. **AGE/SEX:** Most patients are diagnosed in their fifth or sixth decade**CLINICAL PRESENTATION:** Patient presents may with chronic sinusitis, nasal obstruction, pain, non axial displacement of the globe, lower lid swelling, diplopia, blurred vision, epiphora. On examination, patient may have proptosis and displacement of the globe, congestion, chemosis, strabismus, decreased acuity, hypoesthesia, nasal mass. Squamous cell carcinoma is the most common sinus malignancy to involve the orbit these tumors arise from the maxillary sinus in most cases. Nasopharyngeal carcinoma may spread to the orbit through the inferior orbital fissure Retinoblastoma is the most common intra ocular tumor of child hood .The tumor reaches the orbit through the loose connective tissue surrounding the vessels and the nerves .On reaching the orbit, the tumor cells grow quickly as undifferentiated neuroblastic cells without Flexner Winter Steiner rosettes.

TYPES OF SECONDARY ORBITAL TUMOURS:

Paranasal Sinus Tumours: Benign-Mucocele/Osteoma/Inverted papilloma

Malignant-SCC/ADC/ACC/SNUC/Melanoma/Esthesioneuroblastoma

Nasopharyngeal Tumours: SCC/Lymphoepithelioma

Lacrimal Sac Tumours: Epithelial tumours/Melanoma

Conjunctival Tumours: SCC/Melanoma

Eyelid Tumours: BCC/SCC/Seb cell Carcinoma

Intraocular Tumours: Uveal melanoma/Retinoblastoma

IntracranialTumours:Meningioma/Glioblastoma/Chordoma

RADIOLOGY: CT scan of the paranasal sinuses with contrast is particularly important in the initial assessment of sinus malignancies due to the superior capability of the CT scan to demonstrate bony involvement.MRI of the paranasal sinuses with and without gadolinium.MRI also is very useful in determining the extent of sinus disease.MRI can be used to differentiate between inspissated secretions and tumor. MRI is useful in evaluating subtle soft tissue changes. **MANAGEMENT:** According to primary site and type of tumor. Can be wide surgical excision, exenteration, chemotherapy and radiotherapy.

METASTATIC TUMORS[39]

DEFINITION: Invade the orbit by metastasizing from the primary site via heamatogenous or lymphatic spread.**ETIOLOGY:** In adults, breast -42%, lungs -11%, prostate -08%,melanoma-05%, unknown-11%.Paediatric tumors: Embryonal or undifferentiated sarcoma, Neuroblastoma, Ewing's sarcoma, Wilm's tumor. **INCIDENCE:** 10%-12%.**CLINICAL PRESENTAION**: In the child orbit is more frequently involved and the globe less often. Whereas in adults 70% of metastasis are ocular and only 30% are orbital. Orbital metastatic lesion usually presents with an abrupt onset of proptosis, external ophthalmoplegia and orbital pain. Metastases to the orbit most often have indistinct boundaries and are diffusely infiltrating. **RADIOLOGY:** CT shows mass lesion and bone involvement. Osteolytic lesions caused by thyroid and renal cell carcinoma. Osteoblastic lesion caused by prostatic carcinoma.

MANAGEMENT: Observation, systemic hormonal or chemotherapy, radiation,surgical excision, exenteration.

AIMS AND OBJECTIVES

The aims of the study is to

1. Study the demographic distribution of orbital tumors –age wise, sex wise and state wise.
2. Study the presenting complaints, the presenting eye and the duration of the symptoms.
3. Study the history of the associated symptoms and the presence of past and family history
4. Study the clinical features like the visual acuity, puppilary reactions and fundus of the affected eye.

5. Study the laterality of the tumor, type of proptosis, and degree of proptosis.
6. Study the associated clinical features of the affected eye.
7. Study the type of investigative modality used in the diagnosis of the tumor.
8. Study the treatment modality used in the treatment of the tumor.
9. Study the classification of the tumor, the nature of the tumor and type of tumor.
10. To compare the results got with that of international studies.

PATIENTS AND METHODOLOGY

This prospective descriptive study has enrolled patients with orbital tumors, coming to the orbit clinic of a tertiary eye care centre from September 2003 to March 2005.

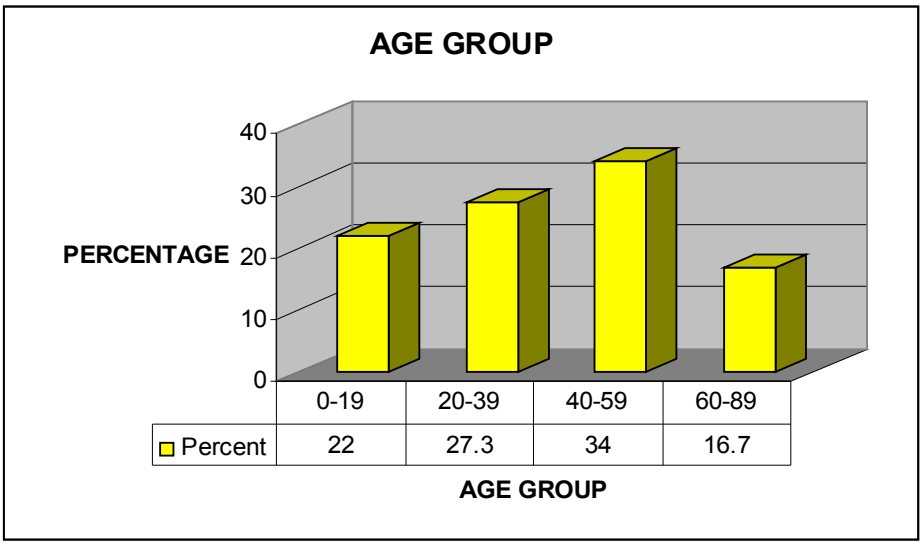
Any case of orbital tumor that presented for the first time to the hospital was included in the study.

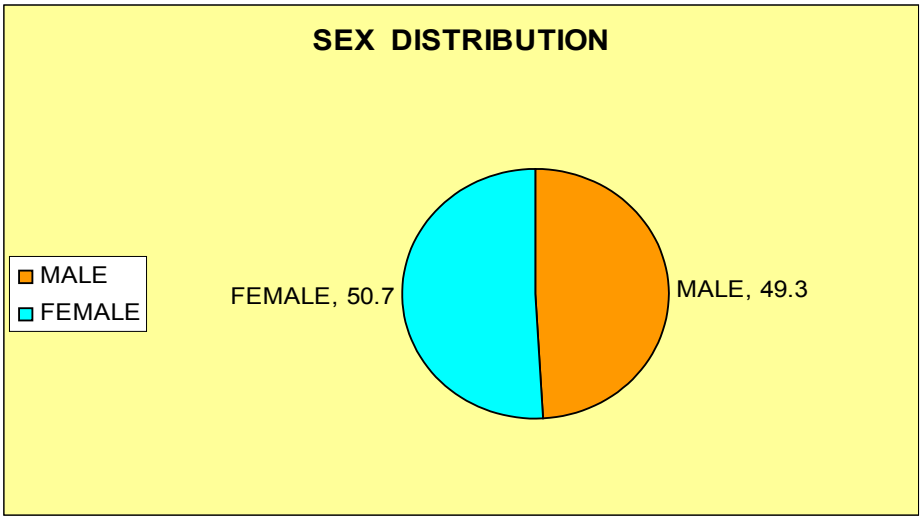
Follow up cases were excluded from the study thereby avoiding repetition.

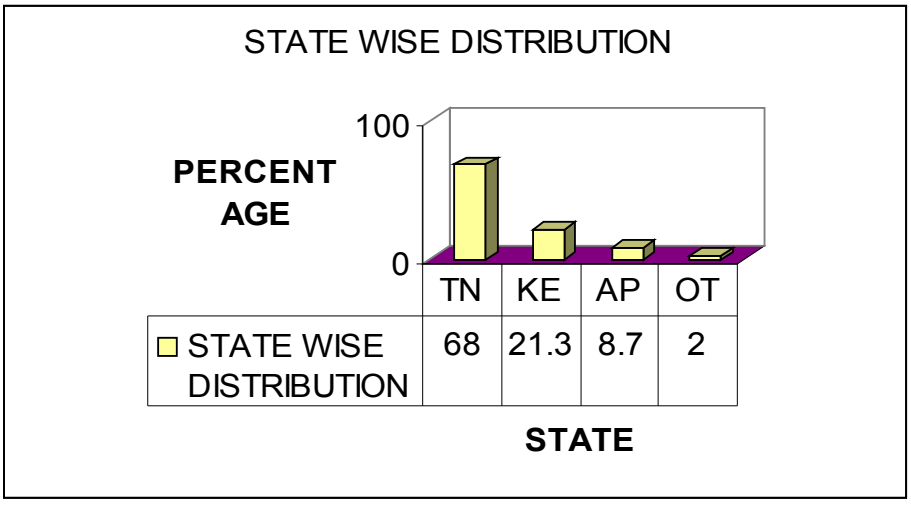
The orbital tumors were diagnosed based on the proforma given below. The diagnosis was based on clinical history and eye examination .Laboratory investigations as well as radiological investigations like X-ray, USG, CT scan were undergone by the patient as required .Histopathological examination using light microscopic examination of H and E stained slides after fixation, paraffin embedding and sectioning were done. Treatment of orbital tumors was also studied.

This study includes orbital tumors diagnosed both by clinical methods and by investigations.

RESULTS


AGE GROUP
PERCENTAGE
40
30
20
10
0
0-19
20-39
40-59
60-89
Percent
22
27.3
34
16.7
AGE GROUP


SEX DISTRIBUTION
MALE
FEMALE
FEMALE, 50.7
MALE, 49.3


STATE WISE DISTRIBUTION
PERCENT AGE
100
0
TN
KE
AP
OT
STATE WISE DISTRIBUTION
68
21.3
8.7
2
STATE

Age Group

	Frequency	Percent
0-19	33	22.0
20-39	41	27.3
40-59	51	34.0
60-89	25	16.7
Total	150	100

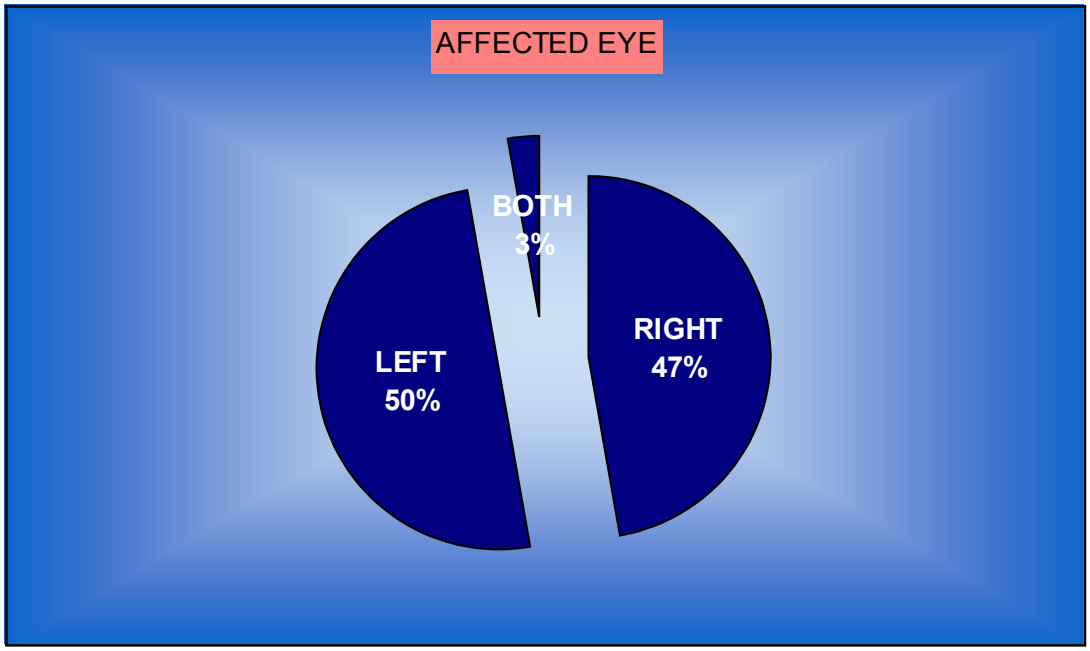
The age wise distribution showed 20%in the 0-19 year age group, 27.3%in 20-39 year age group, 34% in the 40-59 year age group and 16.7% in the 60-89 year age group.

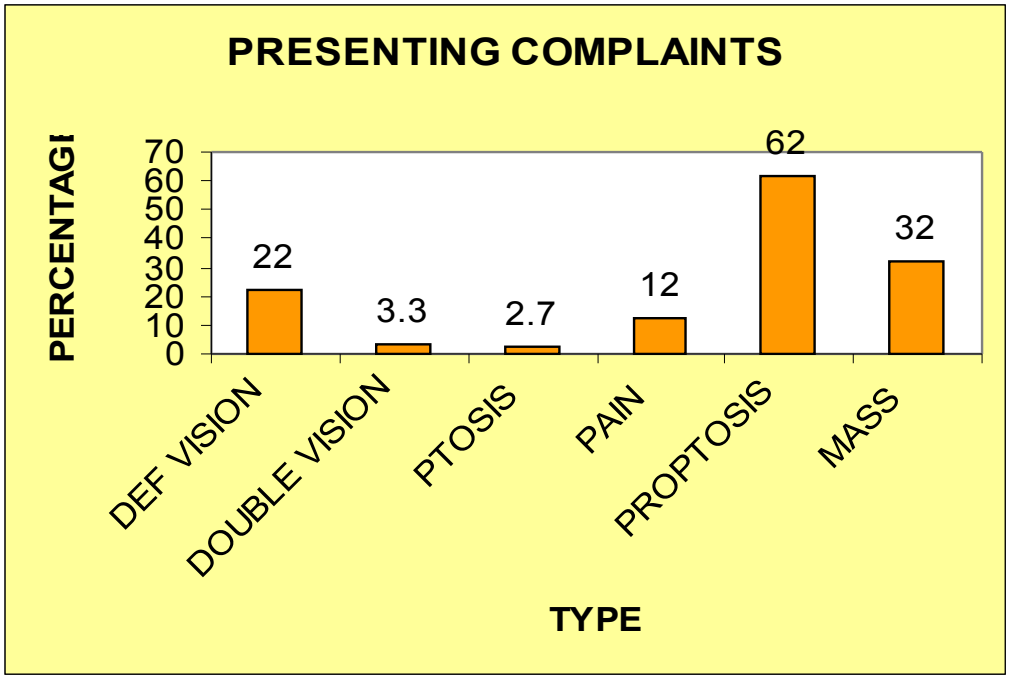
Sex

	Frequency	Percent
Male	74	49.3
Female	76	50.7
Total	150	100.0

The sex wise distribution of orbital tumors showed that in 49.3%of cases males were involved and in 50.7 of cases females were involved.

The state wise distribution of the cases showed that 68% of cases were from Tamilnadu, 21.3% from Kerala, 8.7%from Andra Pradesh and 2% from other states.


AFFECTED EYE
BOTH
3%
LEFT
50%
RIGHT
47%


PRESENTING COMPLAINTS
PERCENTAGE
70
60
50
40
30
20
10
0
22
3.3
2.7
12
62
32
DEF VISION
DOUBLE VISION
PTOSIS
PAIN
PROPTOSIS
MASS
TYPE

Affected Eye

	Frequency	Percent
Right Eye	71	47.3
Left Eye	75	50.0
Both Eye	4	2.7
Total	150	100.0

The affected eye was the right eye in 47.3% of cases and the left eye in 50% of cases .Both the eyes were involved in 2.7% of cases.

Defective vision

	Frequency	Percent
Present	33	22.0
Absent	117	78.0
Total	150	100.0

Double Vision

	Frequency	Percent
Present	5	3.3
Absent	145	96.7
Total	150	100.0

Drooping of eyelids

	Frequency	Percent
Present	4	2.7
Absent	146	97.3
Total	150	100.0

Pain

	Frequency	Percent
Present	18	12.0
Absent	132	88.0
Total	150	100.0

Protrusion of eye ball

	Frequency	Percent
Present	93	62.0
Absent	57	38.0
Total	150	100.0

Mass

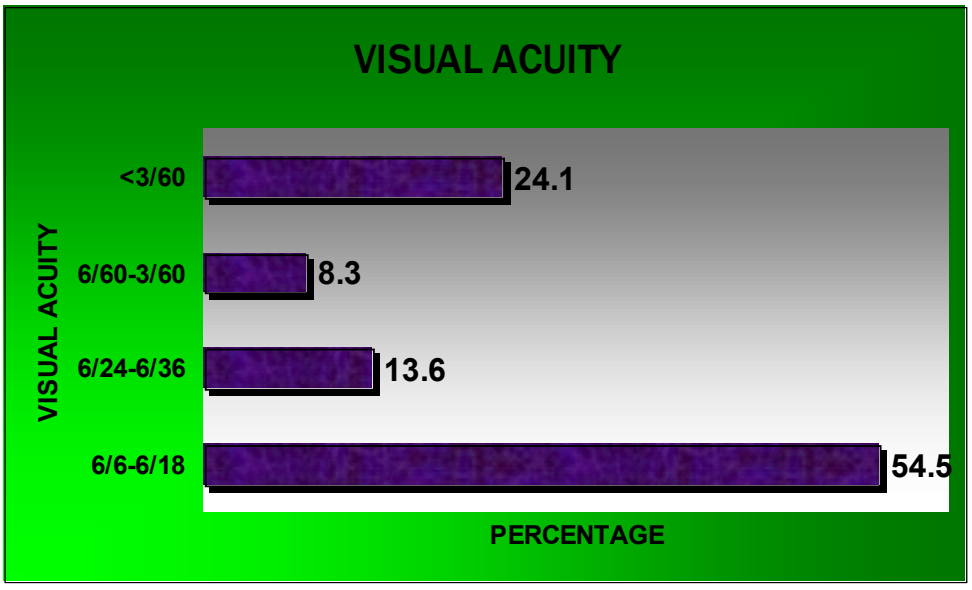
	Frequency	Percent
Present	48	32.0
Absent	102	68.0
Total	150	100.0

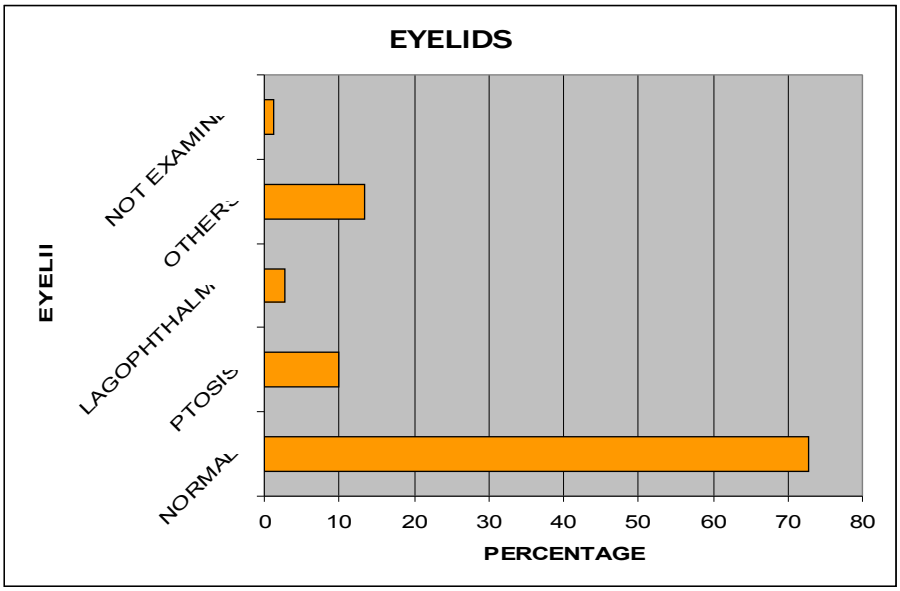
Defective vision was present in 22% of cases, double vision in 3.3%, drooping of eyelids in 2.7%, pain in 12%, proptosis in 62% and a mass was felt in 32 %of cases.

Descriptive Statistics

	N	Minimum	Maximum	Mean	Std. Deviation
Defective vision duration	32	.2	72.0	11.005	16.0309
Double Vision duration	5	1.0	12.0	4.000	4.6368
Drooping of eyelids duration	4	1.0	12.0	6.625	6.2099
Protrusion of eye ball duration	93	.2	120.0	17.147	24.3630
Pain duration	19	1.0	72.0	14.579	25.8442
Mass duration	48	.2	72.0	14.327	16.0925
Valid N (listwise)	0				

The duration of symptoms was a minimum of 2 weeks for defective vision, protrusion of eyeball and mass respectively while it was 1 month for pain, double vision and drooping of eyelids. The maximum duration of symptoms 12 months for double vision and drooping of eyelids, 6 years for defective vision, pain and mass while it was 10 years for protrusion of eyeball.





Group visual acuity

	Frequency	Percent
6/6 – 6/18	79	54.5
6/24–6/36	19	13.1
6/60–3/60	12	8.3
<3/60	35	24.1
TOTAL	145	100

54.5%of patients had visual acuity between 6/6-6/18, 13.1% between 6/24-6/36,8.3% between 6/60-3/60, while 24.1%had vision less than 3/60. 5 patients could not be examined because of their age.

Eye lids

		Frequency	Percent	Valid Percent
Valid	Normal	109	72.7	73.6
	Ptosis	15	10.0	10.1
	Lagophthalmos	4	2.7	2.7
	Others	20	13.3	13.5
	Total	148	98.7	100.0
Missing	System	2	1.3	
Total		150	100.0	

Eye lids were normal in 72.7%of cases while it showed ptosis in 10% of cases, lagophthalmos in 2.7% of cases and other presentation in 13.3% of cases .2 cases could not be examined because one had a squamous cell carcinoma involving the orbit and the other had a basal cell carcinoma involving the orbit.

Conjunctiva

		Frequency	Percent	Valid Percent
Valid	Normal	119	79.3	82.1
	Congested	12	8.0	8.3
	Chemosis	12	8.0	8.3
	Salmon patch	2	1.3	1.4
	Total	145	96.7	100.0
Missing	System	5	3.3	
Total		150	100.0	

Conjunctiva was normal in 79.3% of cases while it was congested in 8% of cases ,chemosed in 8% of cases .Salmon patch was present in 1.3%of cases .5 cases could not be examined because of conjuntival involvement by the tumor.

Pupil

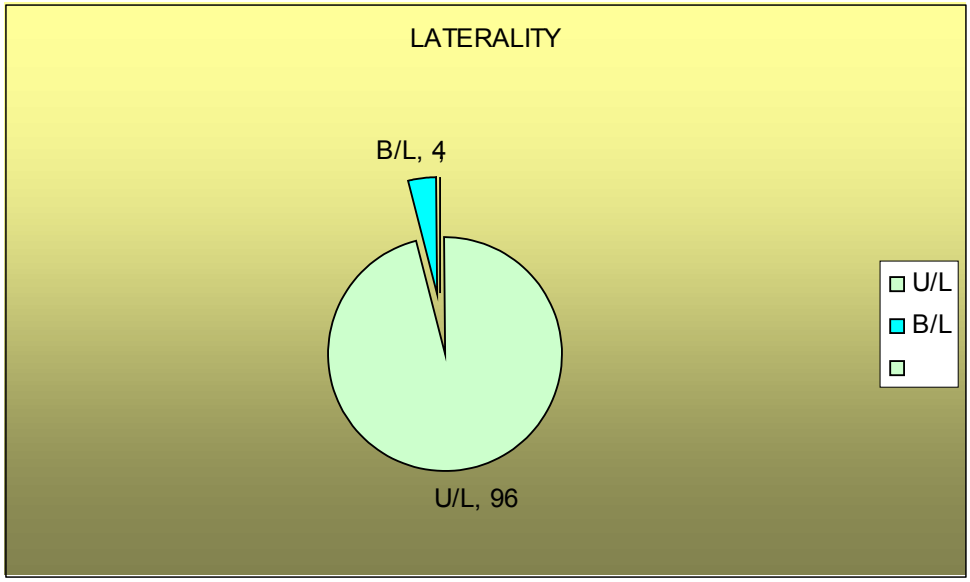
		Frequency	Percent	Valid Percent
Valid	Present	109	72.6	75.7
	Absent	7	4.7	4.9
	RAPD	28	18.7	19.4
	Total	144	96.0	100.0
Missing	System	6	4.0	
Total		150	100.0	

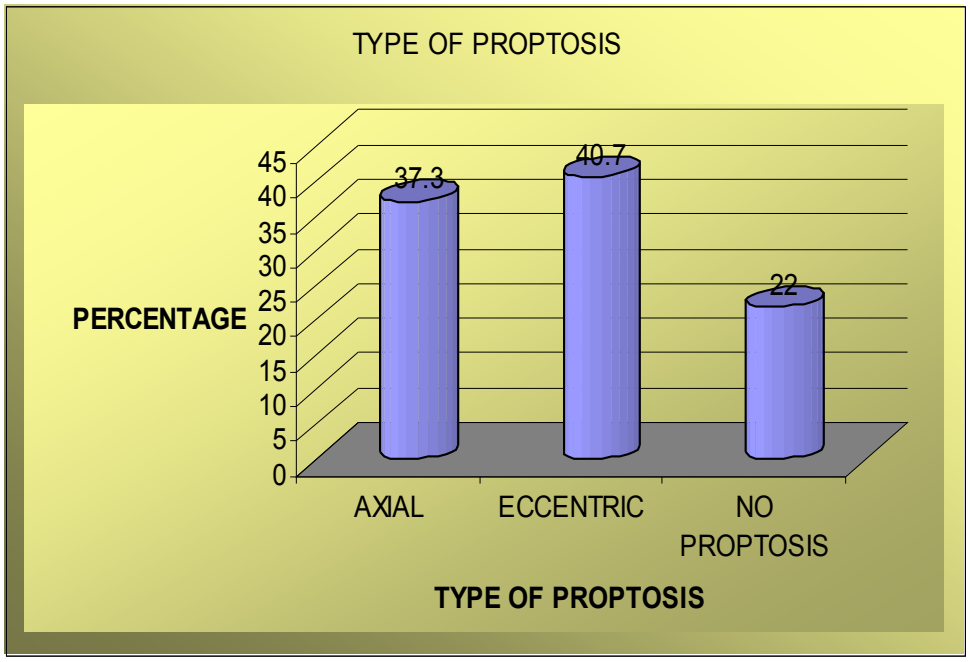
Pupil reaction was normal in 72.7% of cases, absent in 4.7% of cases, RAPD was present in 18.7% of cases.6 cases could not be examined because of the involvement of the cornea by the tumor or the cornea was hazy.

Fundus

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	Normal	107	71.3	76.4	76.4
	Abnormal	33	22.0	23.6	100.0
	Total	140	93.3	100.0	
Missing	System	10	6.7		
Total		150	100.0		

Fundus was normal 71.3% of cases and abnormal in 22% of cases .6.7% of cases could not be examined because of no view of the fundus due to cataract, involvement of the eye by the tumor or hazy cornea.





LATERALITY

	FREQUENCY	PERCENTAGE

UNILATERAL	144	96
BILATERAL	6	4
TOTAL	150	100

Unilateral presentation was in 96% of cases and bilateral in 4% of cases

Globe displacement

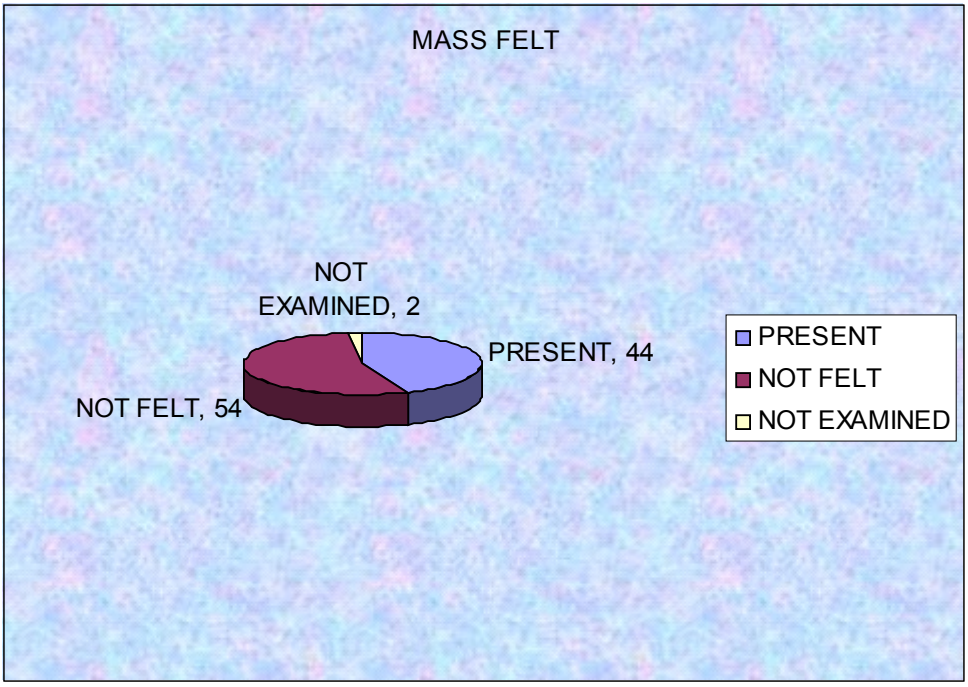
		Frequency		
Valid	Axial	56	37.3	47.9
	Eccentric	61	40.7	52.1
	Total	117	78.0	100.0
NO PROPTOSIS		33	22.0	
Total		150	100.0	

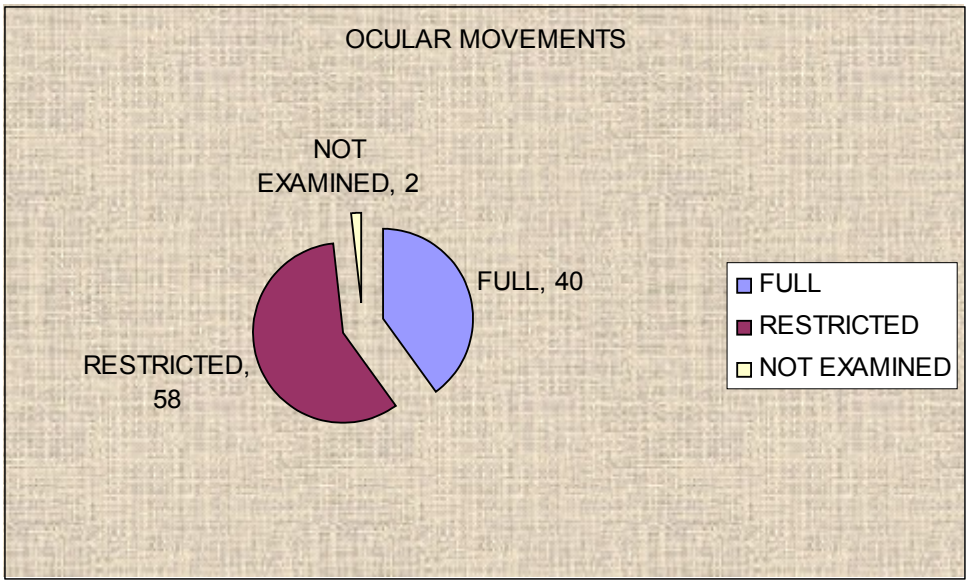
Axial proptosis was present in 37.3% of case and eccentric proptosis in 40.7% of cases. Proptosis was absent in 22% of cases.

Degree of proptosis

	Frequency	Valid Percent
1.00	1	1.8
2.00	6	10.7
3.00	8	14.3
4.00	16	28.6
5.00	2	3.6
6.00	12	21.4
7.00	3	5.4
8.00	3	5.4
9.00	1	1.8
10.00	3	5.4
17.00	1	1.8
Total	56	100.0

Degree of proptosis ranged from a minimum of 1mm to a maximum of 17 mm.


MASS FELT
NOT EXAMINED, 2
PRESENT, 44
NOT FELT, 54
PRESENT
NOT FELT
NOT EXAMINED


OCULAR MOVEMENTS
NOT EXAMINED, 2
FULL, 40
RESTRICTED, 58
FULL
RESTRICTED
NOT EXAMINED

Examination of mass

		Frequency	Percent	Valid Percent
Valid	Mass felt	66	44.0	44.9
	Mass not felt	81	54.0	55.1
	Total	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Warmth

		Frequency	Percent	Valid Percent
Valid	Present	51	34.0	34.9
	Absent	95	63.3	65.1
	Total	146	97.3	100.0
Missing	System	4	2.7	
Total		150	100.0	

Tenderness

		Frequency	Percent	Valid Percent
Valid	Present	18	12.0	12.2
	Absent	129	86.0	87.8
	Total	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Pulsation

		Frequency	Percent	Valid Percent
Valid	Absent	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Finger insinuation

		Frequency	Percent	Valid Percent
Valid	Possible	45	30.0	30.6
	Not Possible	102	68.0	69.4
	Total	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Resistance to retropulsion

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	Present	94	62.7	63.9	63.9
	Absent	53	35.3	36.1	100.0
	Total	147	98.0	100.0	
Missing	System	3	2.0		
Total		150	100.0		

Variation with posture

		Frequency	Percent	Valid Percent
Valid	Present	1	.7	.7
	Absent	146	97.3	99.3
	Total	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Valsalva maneuver

		Frequency	Percent	Valid Percent
Valid	Absent	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Bruit over the mass

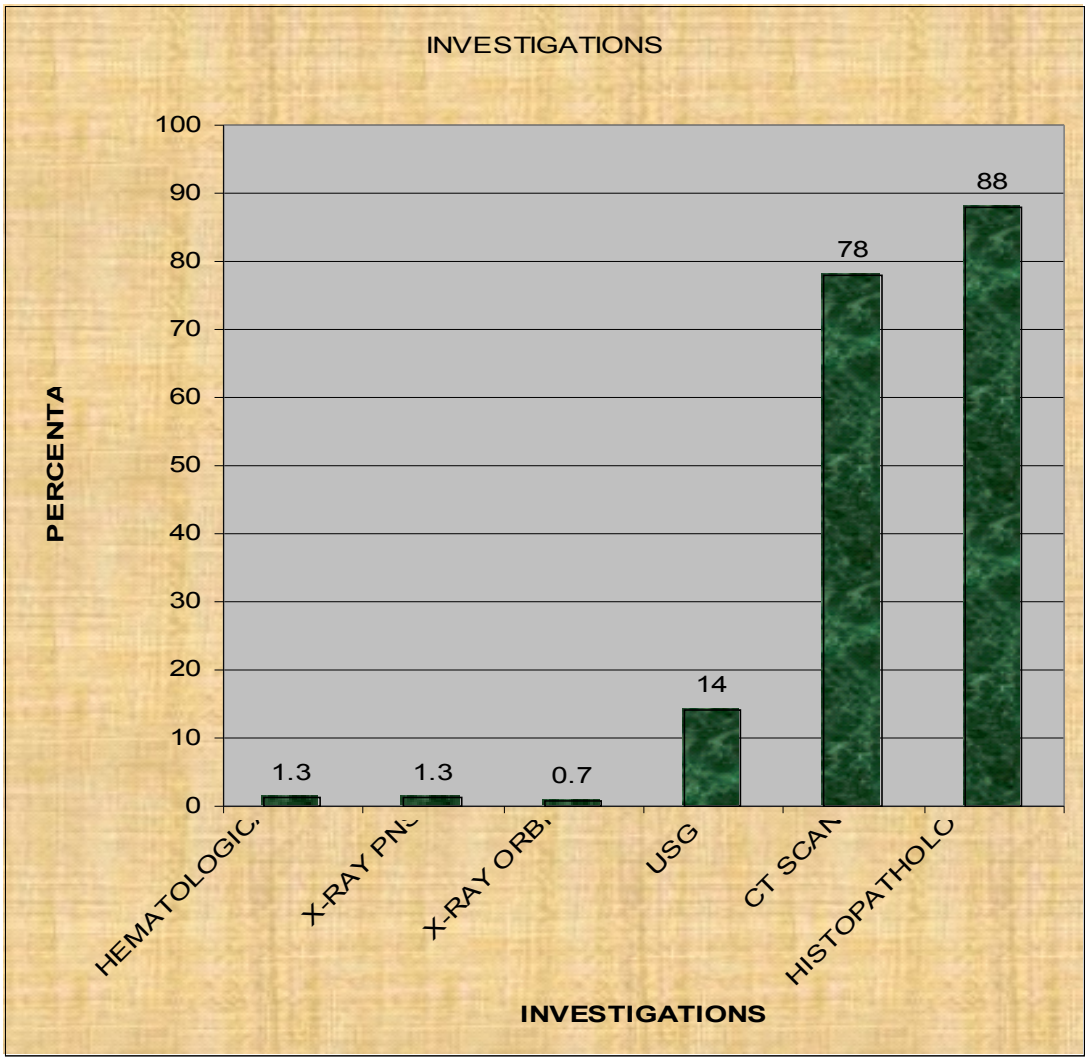
		Frequency	Percent	Valid Percent
Valid	Absent	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Ocular movements

		Frequency	Percent	Valid Percent
Valid	Full	60	40.0	40.8
	Restricted	87	58.0	59.2
	Total	147	98.0	100.0
Missing	System	3	2.0	
Total		150	100.0	

Mass was felt in 44% of cases and was not felt in 54% of cases .2% of cases could not be examined because of involvement of the eyeball by the tumor. Warmth was felt in 34% of cases and was not felt in 63.3% of cases .2.7% of cases could not be examined. Tenderness was present in 12% of cases while it was absent in 86% of cases .2% of cases could not be examined because of involvement by the tumor. Pulsation was absent in all the cases that could be examined (98%).Finger insinuation was possible in 30% of cases and was not possible in 68% of cases. Resistance to retropulsion was present in 62.7% of cases and was absent in 35.3% cases. Variation with posture was present in only.7% of cases and was absent in 97.3% of cases. Variation with valsalva manouevre was absent in all the cases that were examined. Bruit was absent in all the cases examined. Ocular movements were restricted in 58% of cases and were normal in 40% of cases .2% of cases could not be examined because of involvement of the eyeball by the tumor.

INVESTIGATIONS



Haematological

		Frequency	Percent	Valid Percent
Valid	Done	2	1.3	1.3
	Not Done	148	98.7	98.7
	Total	150	100.0	100.0

X-Ray

		Frequency	Percent	Valid Percent
Valid	Not Done	147	98.0	98.0
	PNS	2	1.3	1.3
	ORBIT	1	.7	.7
	Total	150	100.0	100.0

USG B Scan

		Frequency	Percent	Valid Percent
Valid	Done	21	14.0	14.0
	Not Done	129	86.0	86.0
	Total	150	100.0	100.0

Histopathology

		Frequency	Percent	Valid Percent
Valid	Done	132	88.0	88.0
	Not Done	18	12.0	12.0
	Total	150	100.0	100.0

CT Scan

		Frequency	Percent	Valid Percent
Valid	Done	117	78.0	78.0
	Not Done	33	22.0	22.0
	Total	150	100.0	100.0

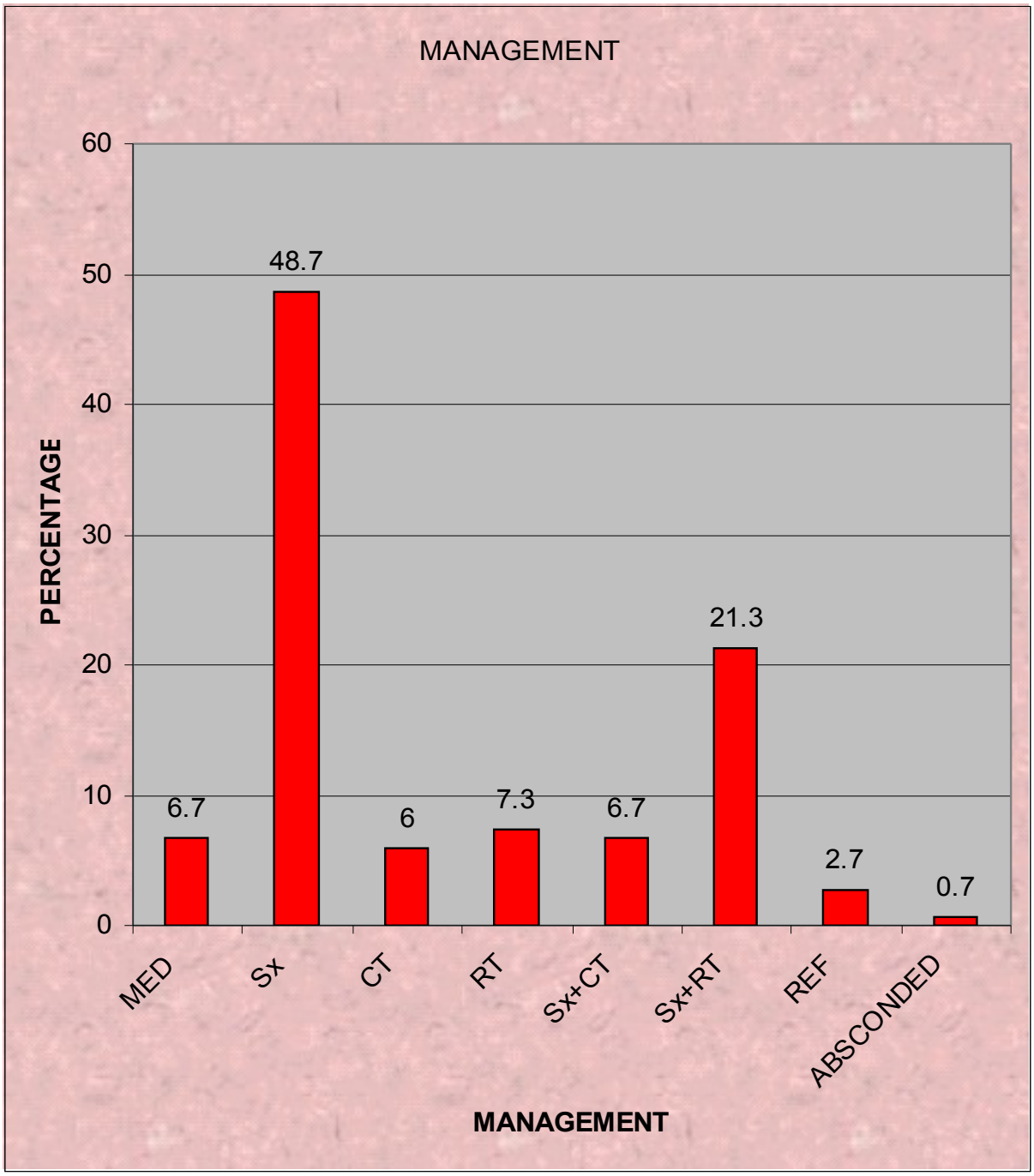
CT Scan

		Frequency	Percent	Valid Percent
Valid	Done	117	78.0	78.0
	Not Done	33	22.0	22.0
	Total	150	100.0	100.0

Histopathology

		Frequency	Percent	Valid Percent
Valid	Done	132	88.0	88.0
	Not Done	18	12.0	12.0
	Total	150	100.0	100.0

Hematological investigation and X-ray PNS was done in 1.3% and X-ray orbits in .7%of cases, USG Scan in 14% of cases, CT scan in 78% of cases and histopathological examination was done in 88% of cases.



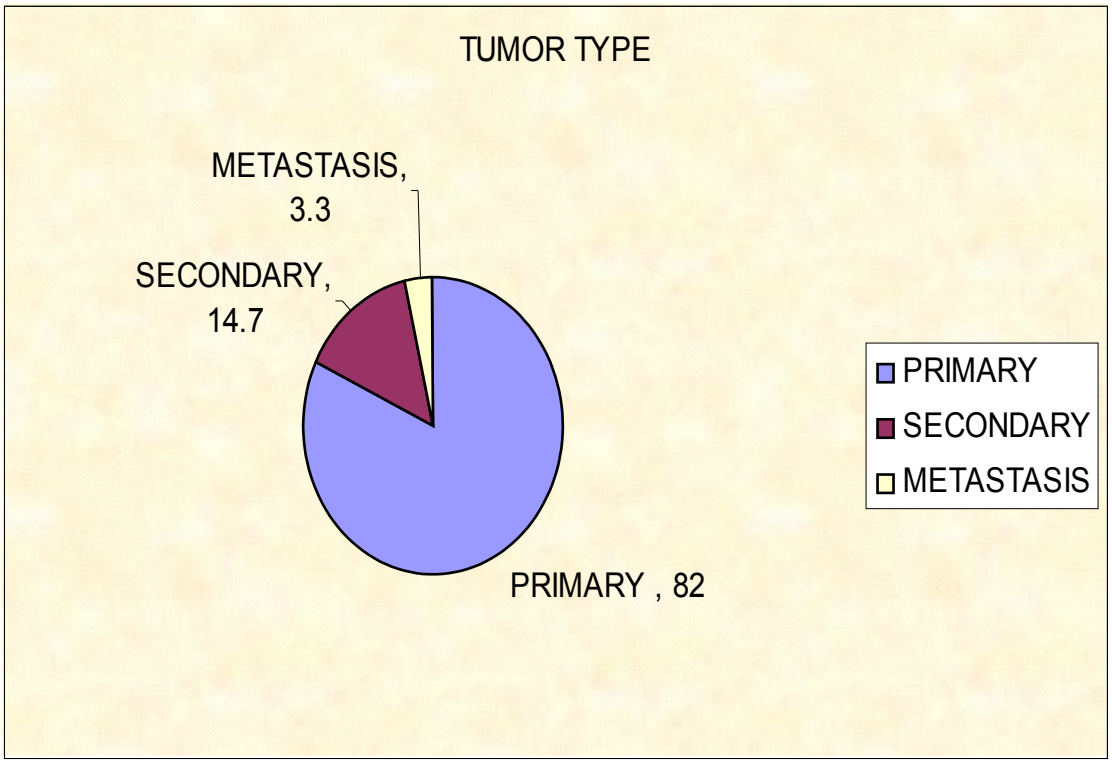
MANAGEMENT

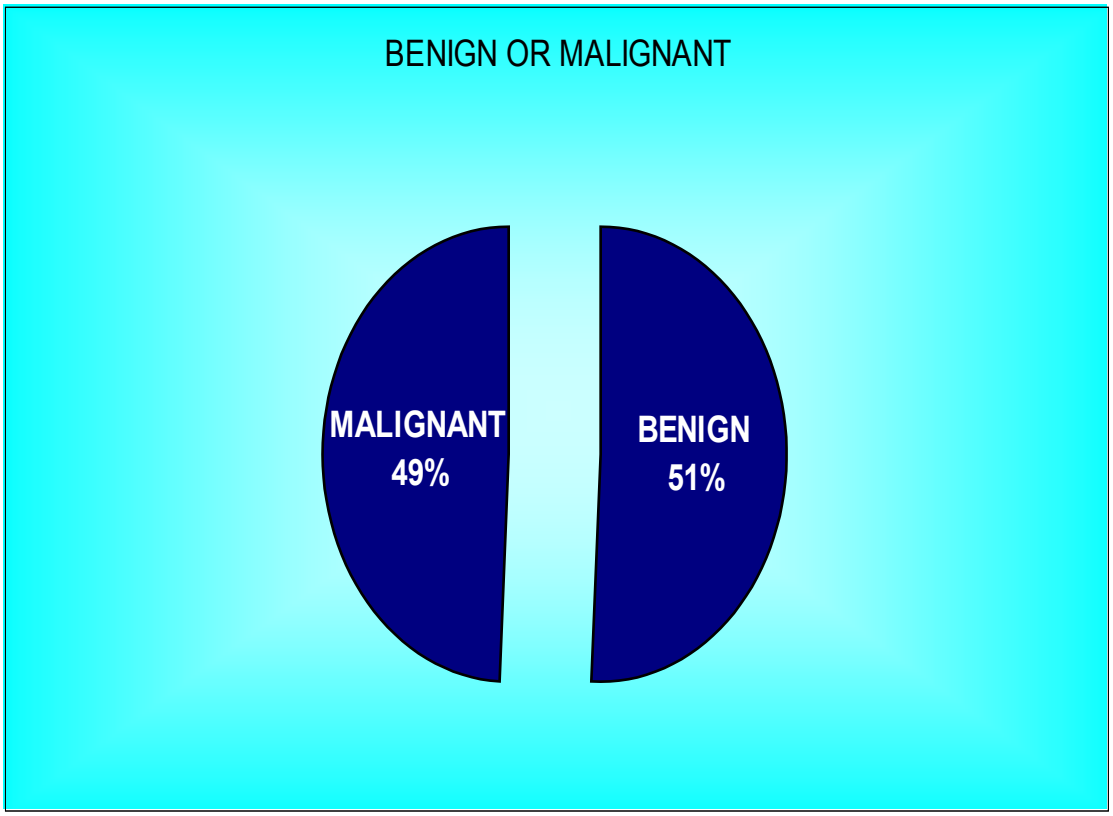
	FREQUENCY	PERCENTAGE

MEDICAL	**10**	**6.7**
SURGERY	**73**	**48.7**
CHEMOTHERAPY	**9**	**6.0**
RADIOTHERAPY	**11**	**7.3**
SX +CT	**10**	**6.7**
SX+RT	**32**	**21.3**
REF	**4**	**2.7**
TOTAL	**149**	**99.3**
ABSCONDED	**1**	**.7**
TOTAL	**150**	**100**

Sx→surgery; CT→chemotherapy; RT →radiotherapy;REF→referred for further management

Medical therapy was instituted in 6.7% of cases. Surgery was done in 48.7% of cases, chemotherapy in 6%, radiotherapy in 7.3%, surgery and hemotherapy in 6.7%, surgery and radiotherapy in 21.3% of cases and 2.7% of the patients were referred. 1 patient absconded.


TUMOR TYPE
METASTASIS, 3.3
SECONDARY, 14.7
PRIMARY , 82
PRIMARY
SECONDARY
METASTASIS


BENIGN OR MALIGNANT
MALIGNANT 49%
BENIGN 51%

DIAGNOSIS

Tumor Type

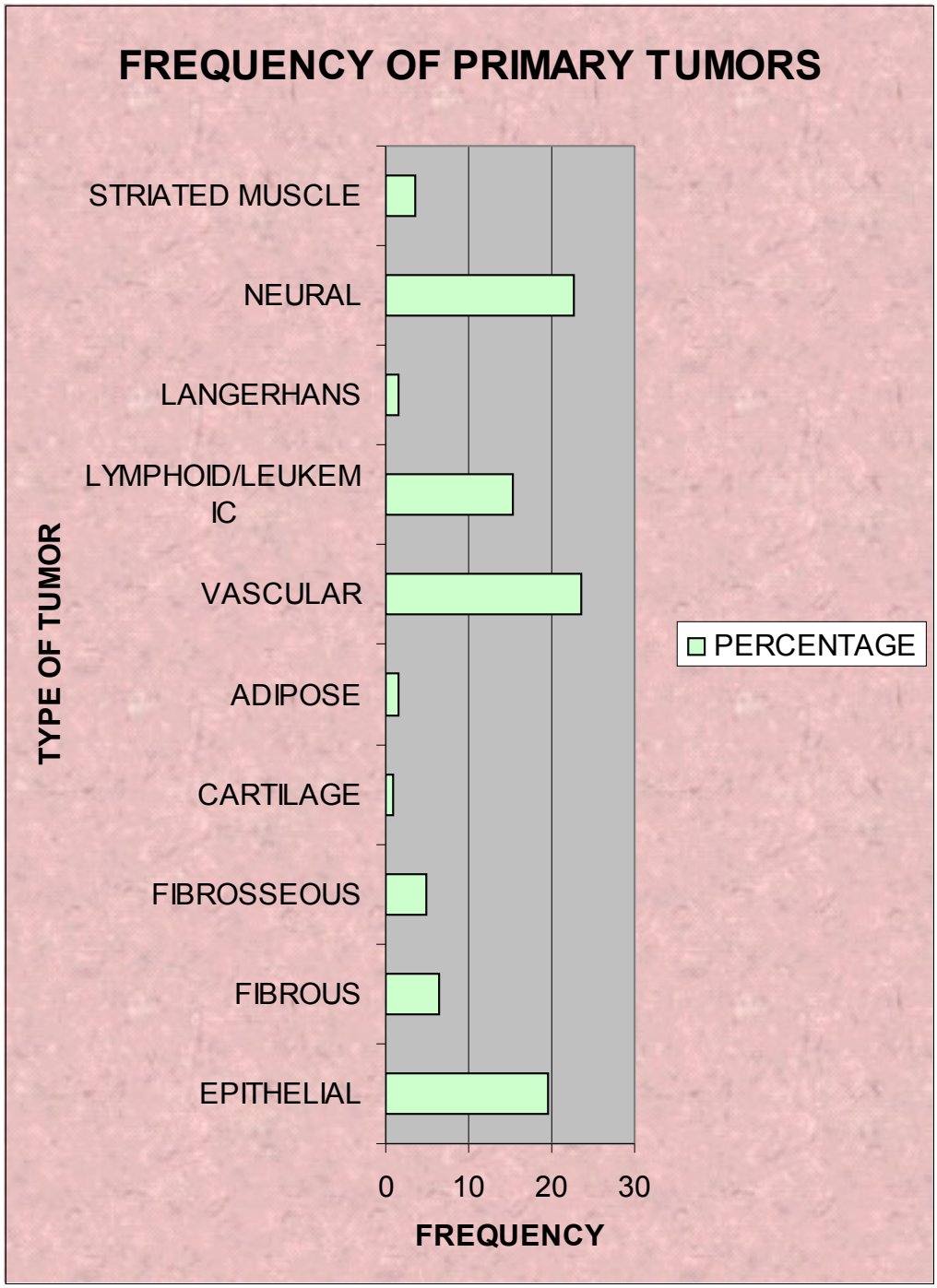
	Frequency	Percent
PRIMARY	123	82.0
SECONDARY	22	14.7
METASTASIS	5	3.3
Total	150	100.0

Primary tumors constituted 82% of the cases, secondary 22% and metastasis constituted 3.3% of cases.

BENIGN OR MALIGNANT

	Frequency	Percent
Benign	76	50.7
Malignant	74	49.3
Total	150	100.0

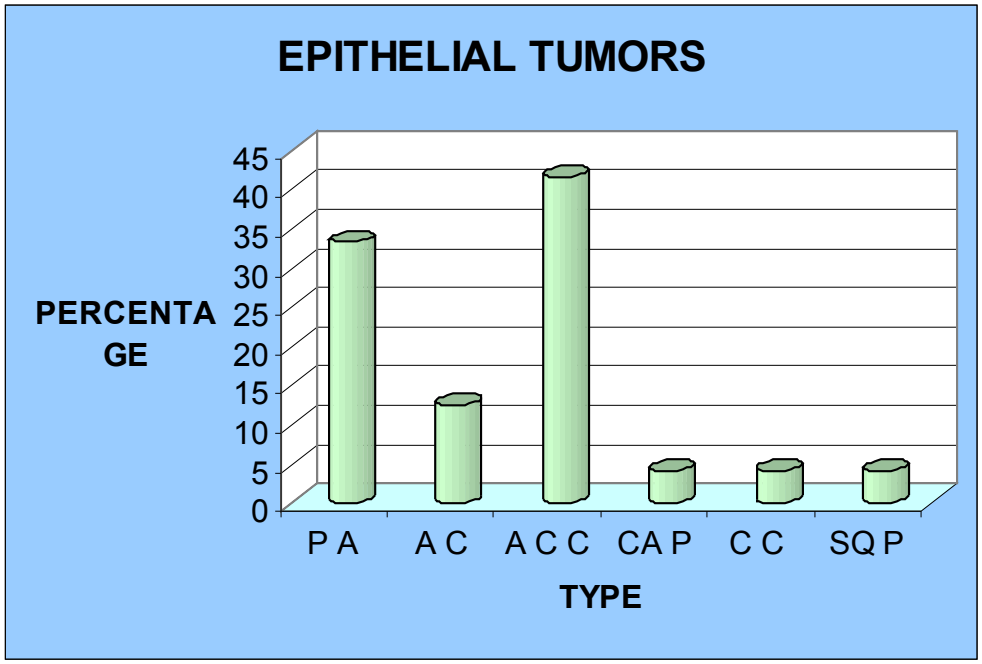
50.7% of the cases were benign tumors and 49.3% of cases were malignant tumors

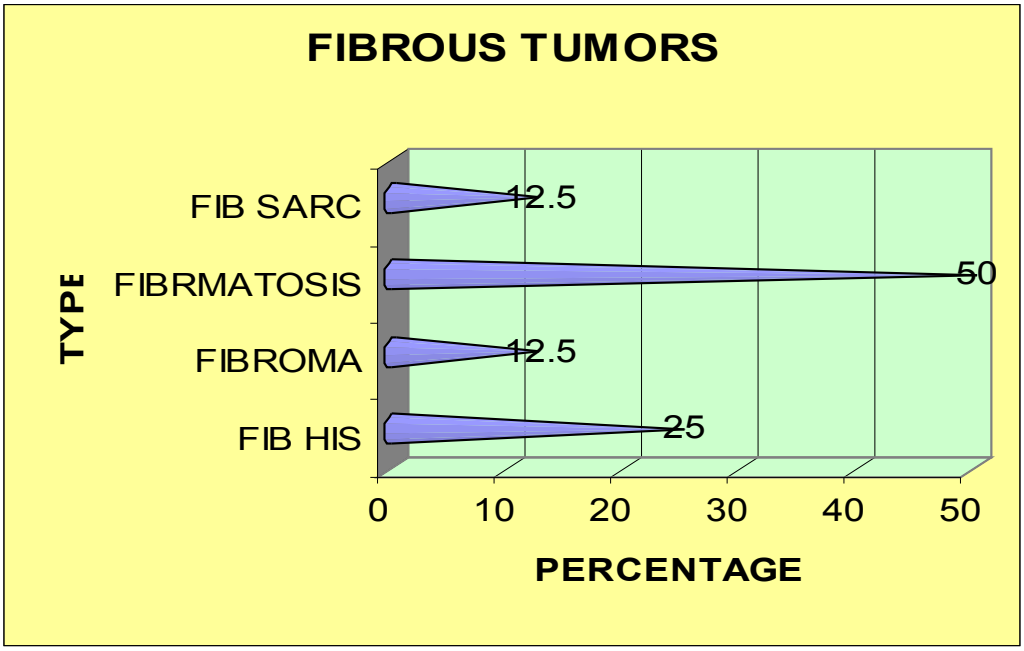

FREQUENCY OF PRIMARY TUMORS
STRIATED MUSCLE
NEURAL
LANGERHANS
LYMPHOID/LEUKEMIC
VASCULAR
ADIPOSE
CARTILAGE
FIBROSSEOUS
FIBROUS
EPITHELIAL
TYPE OF TUMOR
0
10
20
30
FREQUENCY
PERCENTAGE

TUMOR CLASS

	Frequency	Percent
Epithelial	24	19.5
Fibrous	8	6.5
Fibrosseus	6	4.8
Cartilage	1	.8
Adipose	2	1.6
Vascular	29	23.5
Lymphoid/Leukemic	19	15.4
Langerhan's	2	1.6
Neural	28	22.7
Striated muscle	4	3.5
Total	123	100.0

Primary tumors included–epithelial -19.5%, fibrous -6.5%, fibrosseus-4.8%, cartilaginous - .8%, adipose-1.6%, vascular 23.5%, lymphoid and leukemic-15.4%, langerhans cell type -1.6%, neural -22.7% and striated muscle-3.5%.





EPITHELIAL

	FREQUENCY	PERCENTAGE
PLEOMORPHIC ADENOMA	8	33.3

ADENOCARCINOMA	3	12.5
ADENOIDCYSTICCARCINOMA	10	41.6
CA FROM PLEOMORPHIC	1	4.2
CONJUNCTIVAL CYST	1	4.2
SQUAMOUS PAPPILOMA	1	4.2

Pleomorphic adenoma constituted 33.3% of the epithelial tumors,adenocarcinoma12.5%, adenoid cystic carcinoma 41.6%,and carcinoma from pleomorphic adenoma conjunctival cyst and squamous pappiloma being 4.2% each.

FIBROUS

	FREQUENCY	PERCENTAGE
FIBROUS HISTIOCYTOMA	2	25
FIBROMA	1	12.5
FIBROMATOSIS	4	50
FIBROSARCOMA	1	12.5
TOTAL	8	100

Fibromatosis (50%) was the most common tumor of the fibrous variety followed by fibrous histiocytoma (25%) and fibroma and fibrosarcoma being 12.5% each.

FIBROOSSEOUS

	Frequency	Percent
OSSIFYING FIBROMA	4	66.7
OSTEOID OSTEOMA	1	16.7
PAGETS DISEASE	1	16.7
Total	6	100.0

Ossifying fibroma was 66.7%of the osteoid tumors while osteoid osteoma and pagets disease with osteosarcoma was16.7% each.

ADIPOSE

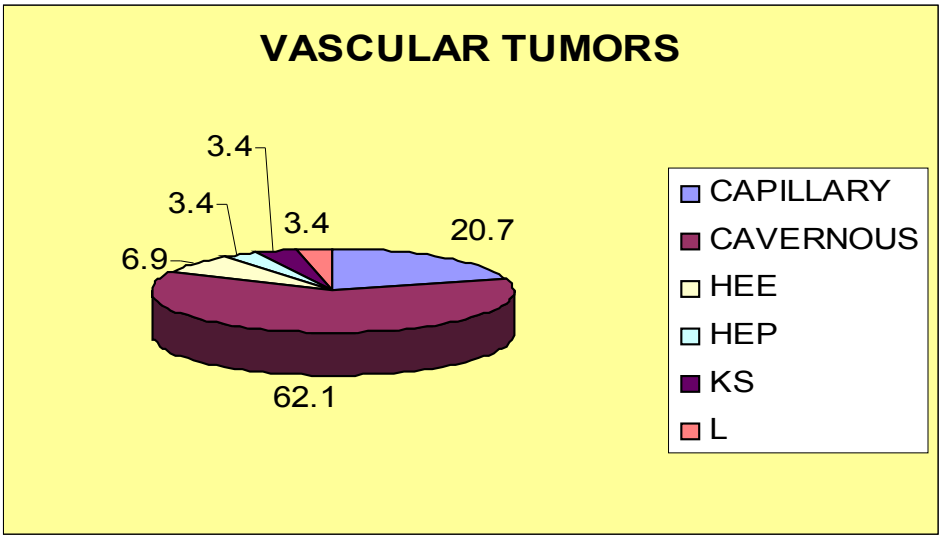
	Frequency	Percent
LIPOMA	2	100.0

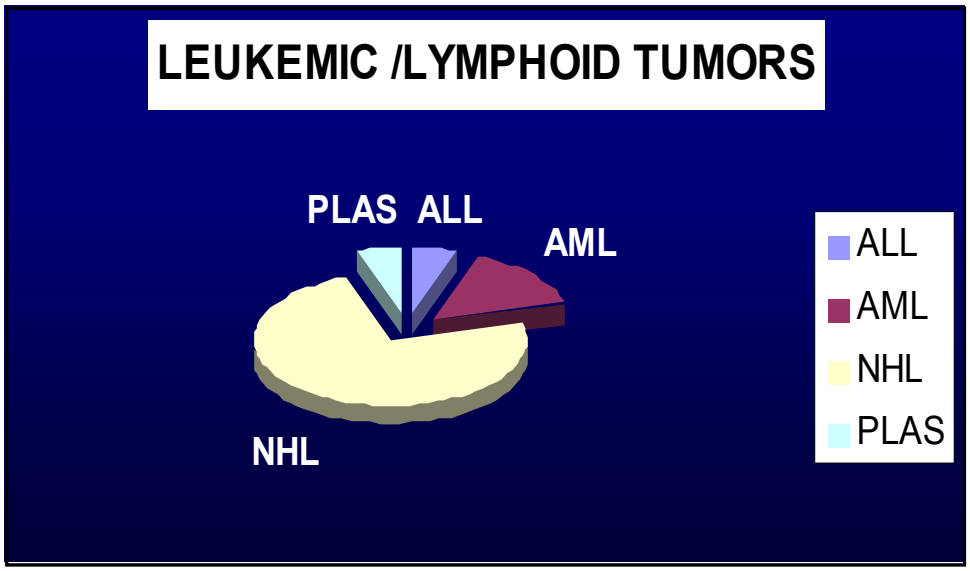
Lipoma constituted 100% of the adipose tissue tumors.

CARTILAGE

	Frequency	Percent
CHONDROSARCOMA	1	100.0

Chondrosarcoma was the only tumor of the cartilaginous variety encountered.





VASCULAR

	Frequency	Percent
CAPILLARY	6	20.7
CAVERNOUS	18	62.1
HEMANGIOENDO	2	6.9
HEMANGIOPERICYTOMA	1	3.4
KAPOSI'S SARCOMA	1	3.4
LYMPHANGIOMA	1	3.4
Total	29	100.0

Cappillary hemangioma constituted 20.7% , cavernous hemangioma 62.1%, hemangioendothelioma6.9%, hemangiopericytoma ,kaposis sarcoma and lymphangioma having3.4% eachof vascular tumors

LYMPHOID /LEUKEMIC

	FREQUENCY	PERCENTAGE

ALL	1	5.3
AML	3	15.8
NHL	14	73.7
PLASMOCYTOMA	1	5.3
TOTAL	19	100

Non Hodgkins lymphoma was the most common leukemic/lymphoid tumor followed by acute myeloid leukemia. Acute lymphoid leukemia and plasmocytoma constituted 5.3% of the cases.

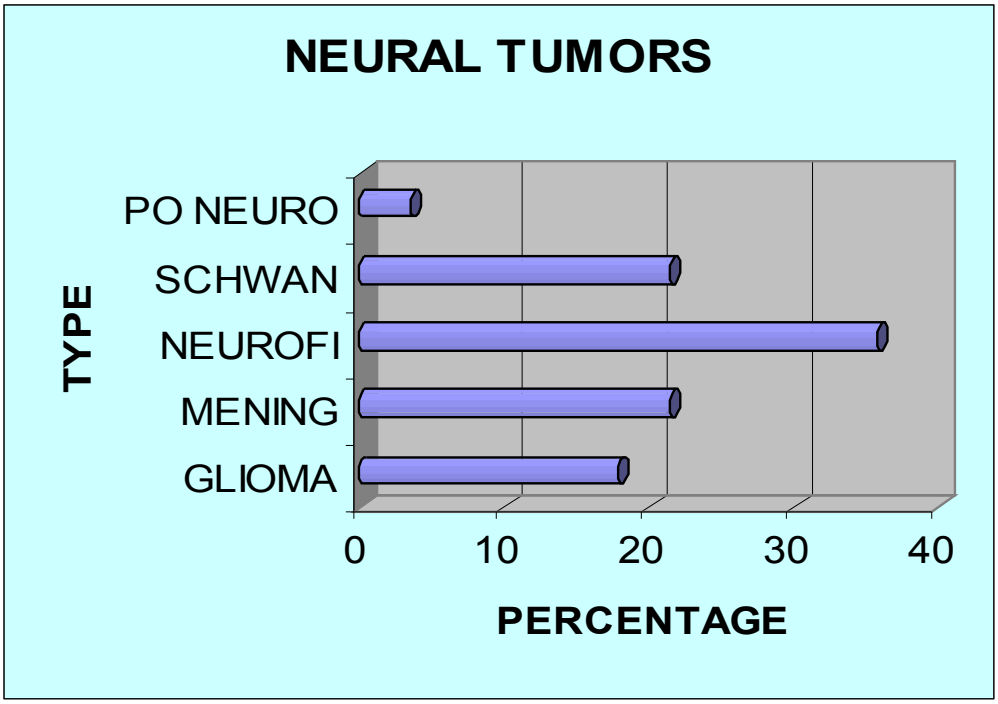
LANGERHANS

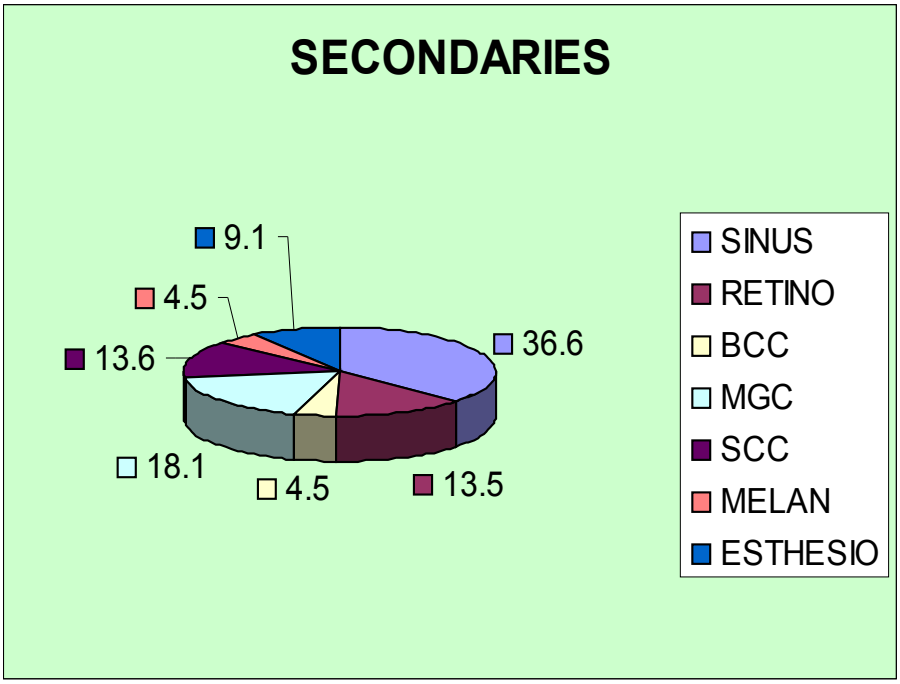
	Frequency	Percent
EOSINOPHILIC	2	100.0
Total	2	100.0

Eosinophilic granuloma was the only tumor encountered in the Langerhans cell variety.

	FREQUENCY	PERCENTAGE
GRANULAR CELL MYOBLASTOMA	1	25
RHABDOMYOSARCOMA	3	75
TOTAL	4	100

Granular cell myoblastoma 25% and rhabdomyosarcoma 75% were the tumors of the striated muscle variety





NEURAL

	FREQUENCY	PERCENTAGE
OPTIC NERVE GLIOMA	5	17.9
MENINGIOMA	6	21.4

SCHWANNOMA	6	21.4
NEUROFIBROMA	10	35.7
PRIMARYORBITALNEUROEPITHI	1	3.6
TOTAL	28	100

Neurofibroma was the most common tumor of the neural variety 35.7%,meningioma and schwannoma being 21.4% each ,glioma 17.9%and primary orbital neuroepithelioma was 3.6%.

SECONDARIES

	FREQUENCY	PERCENTAGE
SINUS MALIGNANCY	8	36.6
BASAL CELL CARCINOMA	1	4.5
MALIGNANT MELANOMA	1	4.5
MEIBOMIAN GLAND CA	4	18.1
ESTHESIONEUROBLASTOMA	2	9.1
RETINOBLASTOMA	3	13.6
SQUAMOUS CELL CA	3	13.6
TOTAL	22	100

Secondaries were common from the sinuses 36.6%,meibomian gland carcinoma 18.1% ,13.6% each retinoblastoma and squamous cell carcinoma esthesioneuroblastoma9.1% and basal cell carcinoma and malignant melanoma 4.5% each.

METASTASIS

	FREQUENCY	PERCENTAGE
PRIMARY UNKNOWN	2	40
BREAST	1	20
CLEAR CELL CA	1	20
CERVIX	1	20
TOTAL	5	100

Metastasis were from breast, cervix and clear cell were encountered but majority of the primary was unknown

DISCUSSION

A spectrum of tumors can involve the orbit .Several publications have addressed the incidence of space occupying lesions in the orbit. However the reported incidence of orbital lesions show great variation .Most reported series of the frequency of orbital tumors are biased by various factors like specialty of the reviewer, histopathological confirmed lesions, age range of patients and geographic area of the patients. The present study differs from the other studies in that it includes patients seen in a tertiary care centre and also on clinical and pathologically proven cases and so reflects the true incidence of orbital tumors seen in ophthalmic practices

Our study is compared to the five major studies done by Shield, Kennedy, Rootman, Henderson and Wilson. Robert Kennedy's series was based on the tumor registry, Shields on the pathology specimens, Henderson summarized his clinical experience, Rootman tabulated his orbital practice and Wilson summarized the orbital lesions submitted to them. This study includes tumors encountered in a tertiary eye care centre and includes both the clinically and the pathologically proven cases.

Kennedy collected his cases in a time period of 34 years, Shields in 20 years, Henderson in 34 years, Rootman in 10 years and Wilson in 51 years while in this study the cases have been collected in a time frame of 18 months.

Type	shields	kennedy	hender	rootman	wilson	Our study
primary	188	284	662	244	76	123

	(69%)	(78.5%)	(63.5%)	(74.8%)	(42%)	(82%)
secondary	70 (26%)	51 (14%)	269 (25.8%)	44 (13.5%)	90 (50%)	22 (14.7%)
metastatic	16 (5%)	27 (7.5%)	111 (10.7%)	38 (11.7%)	15 (8%)	5 (3.3%)

This table shows the distribution of tumors in the six studies compared with each other .Our study shows that the primary tumors were more in number compared to the other studies while secondaries were of same value and metastasis were comparatively less in number

DISTRIBUTION OF PRIMARY TUMORS

Type	Shield	Kennedy	Hender	Rootm	Wilson	Study
Epith	11.1	6.3	10.2	6.1	6.57	19.5
Fibrous	5.85	0.7	3.02	5.3	2.63	6.5
Fibrooseus	4.25	9.85	4.68	8.6	3.9	4.8
Cartilage	1.06	0.35	1.05	0.8	_	0.8
Adipose	1.06	_	1.05	_	3.9	1.6
Vascular	20.2	17.9	18.8	22.9	21	23.5
Neural	12.2	23.5	33.9	29	21	22.7
Striated Muscle	4.25	2.8	5.7	2.04	15.7	3.5
Lymph/Leu k	38.8	37.6	19.3	22.5	21	15.4
Langerhans	0.53	1.05	1.66	1.6	3.9	1.6
Others	1.06	_	0.6	0.8	_	_

Compared to the other studies, more malignant epithelial tumors than pleomorphic adenoma were noted and this may be due to the fact that ours is a tertiary care centre and the malignant lacrimal gland tumors were referred for further management.

In the other studies, fibrous histiocytoma and fibrosarcoma were the most common while fibromatosis was the most common lesion in oour study. All the lesions were confirmed by histopathology.

Ossifying fibroma was the most common tumor in the fibrosseous group which was unlike that of the other studies in which osteoma and fibrous dysplasia were more common .Primary osteosarcoma of orbital bones were rare.

Primary neoplasms in cartilage and adipose tissue were infrequent in all the studies.

Vascular tumors showed capillary and cavernous hemangioma to be the most common tumors .Shields study was based on pathology specimens and so may have underestimated the frequency of these lesions and so may have some basis against these tumors .Our study, as being one including both histological and clinically diagnosed lesions eliminates this basis and hence shows a higher number of vascular tumors.

Optic nerve glioma, neurofibroma and meningioma were the most common tumors of neural origin in all the six studies.

Lymphoid and leukemic tumors were relatively common .The most common lymphoid tumor was Non Hodgkins Lymphoma except in the study done by Shields where lymphoid hyperplasia was more common.

All the tumors of the myogenic variety were malignant in all the six studies.

The three most common primary tumors in all the studies put together were malignant lymphoma, cavernous hemangioma and meningioma while in our study it was malignant lymphoma, cavernous hemangioma, neurofibroma and adenoid cystic carcinoma. All the studies show that the vascular, neural and the lymphoid type of tumors were the most common tumor type. In my study epithelial tumors were found to be more common than in the other studies.

In the secondary neoplasm's invading the orbit the uveal melanoma was found to be the most common ocular tumor invading the orbit while in our study retinoblastoma was the more common one .This may be due to the fact that the uveal melanoma is more common in blue /grey iris than in brown iris. Squamous cell carcinoma was the most common adnexal tumor involving the orbit in the other studies while in our study it was meibomian gland carcinoma. Sinus tumors were also encountered in our study. Other rare tumors like esthesioneuroblastoma were also reported in our study.

In keeping with touch with the prior studies the most frequent metastatic neoplasms have been from the breast .In our study, we had 2 out of the 5 metastatic lesions, in which the primary was unknown

All the studies had different inclusion criteria and the time interval for the six studies were different, which are considered to be the limitations of the study

CONCLUSION

The age wise distribution in the study showed that the most common age group was the 40-59 year age group with the mean age being 39.6 years

In this study females were more commonly involved than males

In this study the most common state from where the patients presented was Tamil nadu and the second being from Kerala

In this study the left eye was more commonly involved than the right eye

The most common presenting symptom was proptosis followed by the presence of a mass. All patients who had the disease before had taken treatment for it before. Family history was positive only in 2% of cases.

The visual acuity was between 6/6 -6/18 in most of the patients

The eye lids and the conjunctiva were normal in most of the cases. The most common presentation in the eyelids were ptosis and the chemosis and congestion and chemosis were present in all the other cases .Pupil reaction was normal in most of the cases and the fundus was normal in most of the cases examined.

Unilateral presentation was more common than bilateral and eccentric proptosis was more common than axial proptosis

Mass felt, warmth ,tenderness ,resistance to retropulsion ,pulsation ,variation with posture and valsalva manouvre and bruit were not felt in most of the cases .Ocular movements were restricted in most of the cases.

Histopathological examination and CT scan were the two most common investigations done

Surgery was the most common modality of treatment followed by a combination of surgery

and radiotherapy.

Benign tumors were more common than the malignant ones.

Vascular, neural and epithelial tumors were the most common tumors encountered.

The most common primary tumors were malignant lymphoma ,cavernous hemangioma ,neurofibroma and adenoid cystic carcinoma.

ANNEXURE

PROFORMA

NAME AGE SEX

MRNO STATE

TAMIL NADU—1
KERALA—2
ANDHARAPRADESH—3
KARNATAKA—4
OTHERS—5

PRESENTING COMPLAINTS

AFFECTED EYE RE→1 LE→ 2 BE→3

SYMPTOMS PRESENT—> 1

ABSENT—> 2

IF PRESENT DURATION

DEFECTIVE VISION

DOUBLE VISION

DROOPING OF EYELIDS

PROTRUSION OF EYEBALL

PAIN

MASS

PAST HISTORY YES→ 1 NO→ 2

IF YES TREATED OR NOT

FAMILY HISTORY YES→1 NO→2

OCULAR EXAMINATION

VISUAL ACUITY RE→ LE→

EYELIDS 1→ NORMAL 2→ PTOSIS 3→LAGOPTHALMOS 4→OTHERS

CONJUNCTIVA 1→NORMAL 2→CONGESTED 3→CHEMOSIS

PUPIL REACTION 1→ PRESENT 2→ABSENT 3→RAPD

FUNDUS 1→NORMAL 2→ABNORMAL

LATERALITY 1→U/L 2→B/L

TYPE OF PROPTOSIS 1→AXIAL 2→ECCENTRIC

IF PROPTOSIS IS AXIAL
HERTLES EXOPHTHALMOMETRY OD→ OS→ BASE→

MASS	PRESENT→1	ABSENT→2
WARMTH	PRESENT→1	ABSENT→2
TENDERNESS	PRESENT→1	ABSENT→2
PULSATIONS	PRESENT→1	ABSENT→2
COMPRESSIBILITY	PRESENT→1	ABSENT→2

FINGER INSINUATION	POSSIBLE→1	NOT POSSIBLE→2
RESISTANCE TO RETROPULSION	PRESENT→1	ABSENT→2
VARIATIONS WITH POSTURE	PRESENT→1	ABSENT→2
VALSALVA MANOUVRE	PRESENT→1	ABSENT→2
BRUIT OVER THE MASS	PRESENT→1	ABSENT→2
OCULAR MOVEMENTS	FULL→1	RESTRICTED→2

INVESTIGATIONS

HEMATOLOGICAL	1→ DONE	2→NOT DONE
X-RAY	1→NOT DONE 2→PNS 3→ORBIT 4→OPTIC FORAMEN 5→SKULL	
CT SCAN	1→DONE	2→NOT DONE
USG	1→DONE	2→NOT DONE
HISTOPATHOLOGY	1→DONE	2→NOT DONE
OTHERS	1→DONE	2→NOT DONE

MANAGEMENT

1→ MEDICAL

2→ SURGERY

3→ CHEMOTHERAPY

4→ RADIOTHERAPY

5→ SURGERY AND CHEMOTHERAPY

6→ SURGERY AND RADIOTHERAPY

7→ REFFERED FOR FURTHER MANAGEMENT

DIAGNOSIS→

TYPE OF TUMOR 1→PRIMARY 2→SECONDARY 3→METASTAS

CLASS OF TUMOR 1-EPITHELIAL 2-FIBROUS 3-FIBROOSSEUS
4-CARTILAGE 5-ADIPOSE 6-VASCULAR
7-LYMPHOID/LEUKEMIC 8-LANGERHANSCELL
9-NEURAL 10-STRIATED MUSCLE
11-SECONDARY 12-METASTATIC

NATURE OF TUMOR 1→BENIGN 2→MALIGNANT

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